Washington State Department of HEALTH

Nutrition Interventions

for Children and Youth with Special Health Care Needs

Fourth Edition, 2024

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Umair Shah, MD, MPH
Secretary of Health

Editors

Sarah Harsh, MS, RDN, CD
Nutritionist
Institute on Human Development & Disability
University of Washington
Seattle, WA

Mari Mazon, MS, RDN, CD
Nutritionist
Institute on Human Development & Disability
University of Washington
Seattle, WA

Merideth Murray, MS, RDN
Founder
Return to Roots Nutrition
Sammamish, WA

Khimberly Schoenacker, RDN, CSP, CD
Nutrition Consultant, Children & Youth with Special Health Care Needs Program, Washington State Department of Health, Olympia, Washington

Authors

Ben Atkinson, MS, RD, CD
Assistant Director, Auburn School District, Auburn, Washington

Ellen K. Bowser, MS, RDN, LDN, RN, FAND
Co-Director and Faculty Nutritionist, University of Florida, Pediatric Pulmonary Division, Gainesville, Florida

Lori Brizee, MS, RD, LD, CDCES
Pediatric Nutritionist, Erika Beard-Irvine MD Pediatrics, Bend, Oregon

Sharon Feucht, MA, RDN, CD
Registered Dietitian/Nutritionist, Retired

Sarah Harsh, MS, RDN, CD
Nutritionist, Institute on Human Development & Disability, University of Washington, Seattle, Washington

continued...

Robin Glass, MS, OTR, IBCLC
Occupational Therapist, Seattle Children's Hospital, Seattle, Washington

Katrina Hoch, PhD, MS, RD
Clinical Dietitian, Seattle Children's Hospital, Seattle, Washington

Kathryn Hunt, RDN, CD
Inpatient Clinical Manager, Seattle Children's Hospital, Seattle, Washington

Anna Joseph, MEd, BCBA, LBA
Clinical Supervisor

Kay Kopp, OTR/L
Occupational Therapist, Institute on Human Development & Disability, University of Washington, Seattle, Washington

Mari Mazon, MS, RDN, CD
Nutritionist, Institute on Human Development & Disability, University of Washington, Seattle, Washington

Deonna Monnahan, MS, RDN, CD, CNSC
Clinical Dietitian, Seattle Children's Hospital, Seattle, Washington

Merideth Murray, MS, RDN
Founder, Return to Roots Nutrition, Sammamish, Washington

Beth Ogata, MS, RDN, CD
Nutritionist, Institute on Human Development & Disability, University of Washington, Seattle, Washington

Annette Pederson, MS, RD, CD
Instructor, Columbia Basin College, Pasco, Washington

Khimberly Schoenacker, RDN, CSP, CD
Nutrition Consultant, Children & Youth with Special Health Care Needs Program, Washington State Department of Health, Olympia, Washington

Sarah Sullivan, MS, RDN, CD
Pediatric Dietitian, Sprouts Nutrition, Charlotte, North Carolina

Valerie Eubanks Tarn, MS, RDN, LDN
Associate Professor of Pediatrics, University of Alabama at Birmingham, Birmingham, Alabama

Umme Salma Vahanvaty, MS, RDN, CSP
Clinical Dietitian, Children's Hospital Los Angeles, Los Angeles, California

Yev Veverka, PhD, BCBA-D
Research Scientist, Haring Center, Seattle, Washington

Nila Williamson, MPH, RDN
Dietitian, Olympia Pediatrics, Olympia, Washington

Lynn Wolf, MOT, OTR, IBCLC
Occupational Therapist, Seattle Children's Hospital, Seattle, Washington

Joan Zerzan, MS
Clinical Dietitian, Retired

Advisory Committee (original edition)

Lori Brizee, MS, RD, LD, CDCES

Lisa Campo, MPH, RD, CD (deceased)

Sharon Feucht, MA, RD, CD

Betty Lucas, MPH, RD, CD

Maria Nardella, MA, RD, CD

Beth Ogata, MS, RD, CSP, CD

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ACKNOWLEDGEMENTS

The 4th edition of *Nutrition Interventions for Children & Youth with Special Health Care Needs* would not have been possible without the dedication and hard work of so many professionals.

Thank you to the authors who contributed their time and expertise to updating this publication. Authors were contacted in late 2019, just months before the COVID-19 pandemic; they made time to research and write their chapters during a very chaotic time. Editors were tasked with reviewing the publication during a time of growing awareness on language and its impact on marginalized communities. We are thankful for their time spent researching inclusive language. Thank you to the registered dietitians who provided recommendations on authors. And of course, the authors who contributed chapters to previous editions of this publication must be acknowledged.

We would like to express gratitude to the Washington State Department of Health's Children and Youth with Special Health Care Needs Program for their continued support of this publication.

We would also like to thank the manager and designers at the University of Washington's Creative Communications Department. This publication would not have been possible without them.

And finally, we would like to acknowledge those who access this publication to provide nutrition services for children and youth with special health care needs.

INTRODUCTION

Maria Nardella and Yuchi Yang
Updated by Mari Mazon

Background

The original edition of *Nutrition Guidelines for Children with Disabilities and Chronic Illnesses* was published in 1989 in response to an assessment of needs for nutrition services in Neurodevelopmental Centers and local health departments throughout the state of Washington.

The primary users in Washington were members of a statewide network of registered dietitians/nutritionists who provide services to children with special health care needs. In 1996, this group was surveyed to determine how useful the book still was as a resource and if there was enough interest and need to warrant a revision. The overwhelming results were to initiate a revision process.

An Advisory Committee was formed and a part time Coordinator/Editor was hired to revise and expand the 1989 version. It was intended to be a contribution to existing tools and nutrition resources for dietetic practitioners that serve to guide or define the provision of nutrition care for children with special health care needs in multiple service settings. It was an expensive endeavor even though hundreds of hours of work were generously donated by the 30 unpaid authors of the various chapters and their employers. It took four years to complete (2001), and the name of the new publication was changed to "*Nutrition Intervention for Children with Special Health Care Needs*." In 2002, a second edition and second printing was done which included minor corrections and additions. The work on the third edition was initiated in 2008 and completed in 2010.

New Edition

Discussion on updating this publication began as we were approaching 10 years since the completion of the 3rd edition. We saw that many national nutrition publications cited this resource, demonstrating its wide use among registered dietitian nutritionists.

Authors were contacted in late 2019, just months before the COVID-19 pandemic. While the chaos and uncertainty during the following 3 years slowed progress, authors generously contributed their time and expertise and made this publication possible.

The title of this publication has been updated to *Children & Youth with Special Health Care Needs*, which acknowledges that youth with special health care needs deserve the same access to

nutrition services as do children. The nutrition interventions included in this resource can be applied to both children and youth.

Organization of the Book

The book is divided into three sections.

Section 1 “Determination of Nutritional Status” outlines the recommended procedures for nutrition screening, and assessment; and addresses the prerequisite steps to take in the development of a nutrition intervention care plan.

Section 2 “Problem-Based Nutrition Interventions” addresses the nutrition-related problems that are more common across a wide range of diagnoses.

Section 3 “Condition-Specific Nutrition Interventions” addresses nutrition management related to specific diseases and disorders that have strong nutrition components.

There is an Appendix that provides more detailed supportive information for the topics presented in the earlier sections and includes many useful tools.

It is hoped that this book will enhance the development of the following skills:

- Comprehensive nutrition assessment
- Nutrition assessment of abnormal growth patterns
- Advanced nutrition assessment and counseling for special diets
- Appropriate uses of special formulas
- Interpretation and application of objective data
- Development of nutrition intervention strategies to produce outcomes
- Participation as a team member to provide interdisciplinary care

Most nutrition and feeding problems of children and youth with special health care needs can be improved, but often are not totally resolved. These children and youth will require ongoing and periodic nutrition assessment and intervention. This book is a resource that will be needed time and again.

INCLUSIVE LANGUAGE STATEMENT

The updated edition of *Nutrition Interventions for Children and Youth with Special Health Care Needs* strives to use inclusive language whenever possible. In 2021, both the American Medical Association and American Academy of Pediatrics published language guides, attesting to the power of words in practice (1,2). The use of inclusive language helps to foster an environment of respect and inclusion for patients and their families and sets the tone for equitable access to health. In this publication, the editors endeavor to reduce biases in the language of health care that may exclude, disrespect, or harm a group of people.

Throughout this text, we have intentionally utilized the following language:

Previous Language	Revised/Inclusive Language	Rationale	Reference(s)
General, non-specific use of pronouns (i.e., he/him, she/her)	They, them, their	The non-specific use of "he" or "she" implies a binary, which excludes individuals identifying as nonbinary. It can also inaccurately gender individuals based on assumptions or stereotypes. The neutral "they/them/their" is used instead. Defer to patient preference in practice.	(1,2,3,4)
Girls and boys	Girls and boys based on sex assigned at birth (context: growth chart genders with sex clarification) -or- Female and male (context: instances where sex assigned at birth is relevant, e.g., energy calculations) -or- Children and youth (preferred term when possible, neutral)	Gender is a social construct, while sex is biologically assigned. Children of a specific sex may not identify with the traditionally assumed, binary genders. Growth curves were developed using participant sex demographics even though the naming of these growth curves includes gender. For clarity, there are instances where we continue to use gendered terms with clarifying language. The neutral "children" and "youth" is used whenever possible.	(1,2,3,4)
Compliance	Adherence	Per the AMA, "compliance" is passive and places sole blame on the patient, whereas "adherence" is more active and considers the numerous influences that affect a person's ability to follow a treatment plan.	(1)
Breast	Breast/chest	"Chest" may be more gender-inclusive, though medically speaking, "breast" is a neutral term. ABM suggests using "mammary gland" in lieu of "breast" (3). In all cases, it is important to ask individuals which terms they prefer in order to provide affirming care.	(5,6)

continued...

Previous Language (continued)	Revised/Inclusive Language	Rationale	Reference(s)
Breast milk	Breast/chest milk, human milk, parent's milk	As in the above, while "breast" may be a neutral term medically, it may also connote female to patients. Terms such as "human milk" and "parent's milk" are more gender-inclusive. Defer to patient/family preference.	(5,6)
Breastfeeding	Breast/chestfeeding	Same as above. ABM also suggests "lactating," "human milk feeding," "expressing," and "pumping" as more inclusive terms (3). Each of these terms has nuance. Defer to patient/family preference.	(5,6)
Mother	Lactating parent, parent	Not all birthing or lactating individuals identify as female or mother. The neutral "lactating parent" or "parent" is used depending on the context. Defer to patient/family preference in practice.	(5,6)
Obese child	Child with obesity	Person-first language is encouraged in health care. Using a disease or condition (such as "obesity/obese") as an adjective can dehumanize patients. It characterizes individuals as their condition, not as a person experiencing their condition. The adjective "obese" should not be used, and the noun "obesity" should be used instead, as in "child with obesity" or "patient with obesity."	(1,2)
Child/children with autism	Child/children with autism (person-first), autistic child (identity-first)	Disability language is evolving, and some groups prefer identity-first language over person-first language. Per the APA, person-first language emphasizes the person over their condition, while identity-first language reflects an intentional reappropriation by an individual of a typically negative term/diagnosis that is embraced as a positive identifier. Always defer to patient preference on whether to use person-first or identity-first language when providing care.	(1,4,7,8,9)
Caucasian, Caucasians	white, white persons	"Caucasian" is an ambiguous racial term, and per the APA, some find it to be offensive. Sources differ in terms of whether "white" should be capitalized or not (e.g., citing the Associated Press, the AMA does not capitalize; APA and CDC do), however, agree that "white," "white persons," or "persons who are white" is preferred over "caucasian." Race is a social construct and should be used as an adjective only. Avoid language that perpetuates negative stereotypes about racial groups. Generally, using a person's geographical place of origin is preferred over race.	(1,3,4)

However, language is dynamic and individuals have unique preferences. The choices we made at the time of publication may not be the norm in the future or may not align with the preferences of the patient and family that you work with. It is important to discuss language with your patients so you can use their preferred language in communication with them.

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CHAPTER 1

NUTRITION SCREENING AND ASSESSMENT

Janet Gilliam, MS, RD, CD and Sandi Laney, RD, CD
Updated 2010 by Roseann Torkelson, MS, RD, CD
Updated 2022 by Sarah Harsh, MS, RDN, CD

Nutrition disorders and compromised nutritional status commonly occur among children and youth with special health care needs. A survey of children from birth to age three years with developmental delays in early intervention programs found 70-90% had one or more nutrition risk indicators (2). Indicators of nutritional risk include altered growth, increased or decreased energy needs, medication-nutrient interactions, metabolic disorders, impaired ability to utilize nutrients, poor feeding skills, and partial or total dependence on enteral or parenteral nutrition (1). A well-nourished child has increased alertness and stamina to participate in therapies, educational activities, and social interactions and benefits from fewer illnesses and improved coping skills. Improved nutritional status and feeding skills may increase the level of independence the child is able to achieve. It can improve the child's perception of self and the caregivers' perceptions of their abilities to meet the child's needs (2).

Screening and assessment of nutritional status are integral components of pediatric health care (3). Screening is a preliminary survey of factors associated with nutritional status that is undertaken to identify children and youth who appear to have nutrition problems or who are at risk for developing a nutrition problem (4). Nutrition screening should be routinely performed for all children and youth with special health care needs. Screening provides general information that can be used in the more comprehensive Nutrition Care Process of nutrition assessment and diagnosis, leading to nutrition intervention, monitoring, and evaluation (5).

Nutrition Screening

Nutrition screening has a variety of functions, requirements, and benefits. Screening consists of the collection of preliminary data in one or more of the following categories:

- anthropometric parameters
- clinical (medical history and diagnosis)
- biochemical laboratory data
- behavior (related to feeding)
- diet

- developmental feeding skills
- socioeconomic characteristics

The screening activities in each of these categories are described in Table 1−1.

Nutrition screening can be effective without including all categories or all suggested data within a category. The screening protocols must be adapted to the setting and according to staff availability and other resources (6). Nutrition screening should be brief and easy to administer. Parent-administered questionnaires and/or interview methods can be effective tools for obtaining screening data. Screening can be successfully completed by a variety of individuals such as the parent or caregiver, public health nurse (PHN), clinic nurse, therapist, social worker, family resource coordinator (FRC), primary care provider (PCP), registered dietitian nutritionist (RDN), or dietetic technician (DTR). Nutrition screening can be incorporated into initial early intervention screenings so that concerns can be identified and children can be referred for an assessment. Children and youth need to be screened on a regular basis to monitor growth and nutritional status over time. Sample screening forms are included in Appendix A.

When a child is identified as having one or more nutritional risk indicators, referral for nutrition assessment with an RDN is needed. Nutrition risk indicators need to be clearly defined to avoid over-identification or under-identification of those at risk. Table 1-1 includes examples of risk indicators and sample criteria. Actual criteria used should be determined individually to reflect the setting and population served for each environment where screening will be done. In addition to red flags identified by nutritional risk indicators, parental concerns should be carefully listened to and considered.

Nutrition Assessment

Once a nutritional risk indicator is identified through screening, a nutrition assessment serves to obtain all information needed to rule out or confirm a nutrition-related problem. Nutrition assessments should be completed by an RDN, preferably with pediatric expertise and/or specialized training for children and youth with special health care needs and developmental disabilities.

Nutrition assessment consists of an in-depth and detailed collection and evaluation of data in the following areas: anthropometrics, physical exam, clinical/medical history, diet, developmental feeding skills, behavior related to feeding, and biochemical laboratory data (2). During the assessment, risk factors identified during nutrition screening are further evaluated and a nutrition diagnosis can be made. The assessment may also reveal areas of concern such as oral-motor development or behavioral issues that require referral for evaluation by the appropriate therapist or specialist. The nutrition assessment is one of the essential elements of a comprehensive interdisciplinary team evaluation and intervention plan. Table 1-2 provides parameters for

completing nutrition assessments and example indicators for nutrition intervention. The indicators used should depend on the specific patient and environment.

Nutrition Intervention

Planning and providing nutrition care and intervention for children and youth with special health care needs is often complex because many factors interact to affect nutritional status. Optimal nutrition care involves consultation and care coordination with professionals from a variety of disciplines. The interdisciplinary team may consist of the child and family, PCP, occupational therapist (OT), physical therapist (PT), speech language pathologist/therapist (SLP), RDN, behavior specialist, social worker, PHN, and home health care providers. Other community agencies such as schools, early intervention programs, hospitals, specialty clinics, the Special Supplemental Nutrition Program for Women, Infants and Children (WIC), Head Start, day care, Division of Developmental Disabilities, and Child Protective Services may also be involved.

The team approach consists of professionals working in a family-centered partnership to coordinate services and provide continuity of care for the child and family. With input from team members, a specific plan of nutrition intervention is developed. The nutrition intervention step of the Nutrition Care Process should have a preventive emphasis and take into account the cultural norms and traditions of the patient and their family. Nutrition care goals and objectives (or outcomes) can become a part of the child's Individual Education Plan (IEP) or Individualized Family Service Plan (IFSP) (See Chapter 12). Reassessment should occur at regular intervals to monitor the child's nutrition status and evaluate the effectiveness of the nutrition intervention. Based on the reassessment, nutrition goals and objectives may be modified to meet the needs of the child and family (5).

Table 1-1: Nutrition Screening [3—7]

Repeat screening in 6 to 12 months if no nutritional risk factors are identified.

Screening Activities	Example Nutrition Risk Indicators
Anthropometric Measure and weigh using standardized techniques and appropriate equipment[1]. Plot on standard growth charts[2]. • Height or length for age • Weight for age • Weight for height (or length) • Head circumference (under age 2 years) • Body Mass Index (BMI) (over age 2 years) Compare current measurements to reference data and to previous measurements available.	Refer for nutrition assessment if any of the following exist: • Height or length for age: z-score <-1 • Weight for age: z-score <-1 • Weight for length (or height): z-score <-1 • Weight for length (or height): greater than 90th percentile • BMI: greater than 85th percentile or z-score <-1 • Change in weight or length of 2 or more percentile channels or a change in z-score by 1.0 or more • Inadequate growth or weight gain for more than one month (under age two)
When doing anthropometrics, observe for signs of neglect or physical abuse.	If signs of neglect or physical abuse are noted, contact Child Protective Services (CPS).
Biochemical Laboratory Data Obtain lab data from medical record, WIC program, or primary care provider: • Hematocrit (Hct) or hemoglobin (Hgb) • Other pertinent lab data: ferritin, vitamin D, if available	Refer for nutrition assessment if abnormal lab values of nutritional significance.
Clinical/Medical History Review past medical history and current health status and diagnosis	Refer for nutrition assessment if any of the following: • Anemia • Anorexia and/or bulimia nervosa • Autism spectrum disorder (See Chapter 21) • Cardiac, pulmonary, or renal disease (See Chapters 15 and 17) • Chronic constipation or diarrhea (See Chapters 6 and 7) • Chronic diseases such as diabetes, cancer, HIV/AIDS • Cystic fibrosis (See Chapter 16)
Screening Activities	**Example Nutrition Risk Indicators** • Feeding problems, poor appetite, or refusal to eat (See Chapters 8 and 9) • Fetal alcohol syndrome or fetal alcohol effects • Food allergies or intolerances • Frequent or recurring infections • Gastrointestinal disorders, reflux, vomiting • History of poor growth or excessive weight gain (See Chapter 13) • Long-term use of laxative, diuretic, anticonvulsant, steroid, or stimulant medications (See Chapter 5) • Malabsorption syndromes • Metabolic disorders, i.e., PKU, galactosemia (See Chapter 19) • Myelomeningocele (spina bifida) • Neurological conditions, i.e., cerebral palsy, anoxia, trauma • Oral or facial anomalies that affect nutrition (See Chapter 8) • Prader-Willi syndrome (See Chapter 13) • Significant dental problems • Special or therapeutic diet (See Chapter 19) • Tube feeding (See Chapter 10)

Dietary Interview caregiver(s) to determine • Concerns about food intake, feeding, and nutrition • Child's typical feeding pattern (e.g., types of foods eaten and how often, aversions, and preferences) • Use of oral supplements • Use of vitamin/mineral supplements • Use of herbal products, alternative nutrition, or other therapies	Refer for nutrition assessment if: • Inadequate or inappropriate dietary intake, i.e., NPO or hypocaloric intake for more than 3 days • Alternative or special diet: vegan, gluten free, multiple food allergies, or other restricted diet • Consumes only liquid, pureed, or ground food after age 2 • Pica (intake of non-food items, i.e., clay, dirt, starch) • Use of supplements, including vitamin/ minerals exceeding 100% of the RDA without physician recommendation
Developmental Feeding Skills[3] Interview caregiver(s) to determine child's feeding skills: • Oral-motor control • Frequency and duration of feedings • Consistency of foods eaten • Self-feeding skills • Typical fluid intake by breast, bottle, and/ or cup • Concerns about progression of feeding skills Review health records for signs of delays or abnormalities in the development of feeding skills.	Refer for nutrition assessment if: • Abnormal sucking pattern (arrhythmic, disorganized, lack of initiation) • Swallowing difficulties (gagging, choking, coughing, noisy breathing after feeding) • Difficulty with chewing • Inability to drink from a cup at appropriate age • Lack of progression in food textures • Not self-feeding after two years of age • Feeding routinely takes longer than 45 minutes per meal
Interview caregiver(s) about child's behavior during feeding. Review health records for signs of behavior problems related to feeding.	Refer for nutrition assessment if signs of behavior problems related to feeding, including: • Disruptive behavior at mealtime • Refusal to eat • Voluntary gagging on foods
Socioeconomic Characteristics Obtain by interview or review of health records: • Family size and income level • Cultural and familial food patterns • Adequacy of food resources • Participation in food and community programs: WIC, SNAP, school food program, food banks	Refer for nutrition assessment if inadequate or inappropriate food pattern: • Insecure food supply • Inadequate housing • Abusive home situation • Financial difficulties Refer to appropriate social services, nutrition programs, and/or food resources.

[1] See Chapter 2.
[2] Correct for prematurity up to age 2 years. See Chapters 2 and 14.
[3] See Chapter 8 for appropriate developmental milestones.

Table 1-2: Nutrition Assessment [4,5,7—11]

Assessment Activities	Example Indicators for Nutrition Intervention
Anthropometric[4] Measure and weigh using standardized techniques and appropriate equipment. For difficult to measure children, arm span, crown-rump, or sitting height may be appropriate methods to estimate stature. Plot on WHO or CDC Growth Charts. Use specialized growth charts as indicated[5]: • Height or length for age • Weight for age • Weight for height (or length) • Head Circumference (under 2 years) • Body Mass Index (BMI) (over age 2 years) Measure and calculate, if skilled in these techniques: • Mid-upper arm circumference (MUAC) • Triceps skinfold • Subscapcular skinfold • Arm muscle area • Arm fat area Compare all current measurements to reference data and previous measurements.	Nutrition intervention may be indicated if any of the following: • A decline in z-score for any anthropometric measure. • Increase in weight gain velocity that exceeds an increase in length/height gain velocity • Any anthropometric measure consistent with malnutrition. See Table 1-3.
Nutrition Focused Physical Exam The following are the components and typical findings of a nutrition focused physical exam. Variations from the findings below may be a sign of nutrient deficiency or excess. • Skin examination- elastic and firm without lesions, rashes, or hyperpigmentation • Hair- shiny, firm, and elastic • Nails- convex and without ridges • Head and neck- symmetric facial features and evenly molded head without obvious thyroid gland in neck • Eye- clear, bright, and shiny • Lip- smooth and moist	Nutrition intervention may be indicated if the findings from the nutrition focused physical exam are confirmed with biomedical data. Refer to primary care for testing. If signs of neglect or physical abuse are noted, contact Child Protective Services (CPS).
• Mouth- deep pink tongue without edema with firm, coral color gums and white smooth teeth without spots or pits. • Cardiovascular- ask about abnormalities in blood pressure, pulse, or arrhythmias • GI- note constipation or diarrhea • Musculoskeletal- look for atrophied muscles, edema, or non-symmetric or bowed extremities. Could consider using grip strength measurement • Neurological- alert and responsive without unsteadiness • Sexual Maturation- note variation from age appropriate sexual development When doing anthropometric measurements or nutrition focused physical exam, observe for signs of neglect or physical abuse. See Reference 7, Table 4-18 and Pocket Guide to Pediatric assessment, Chapter 6	

[4] See Chapter 2.
[5] Correct for prematurity up to age 2 years. See Chapters 2 and 14.

Biochemical Laboratory Data Recommend or obtain the following lab tests as indicated by anthropometric, clinical, and dietary data. Consult with child's primary care provider or clinic physician for appropriate tests. • Complete blood count (CBC) • Tests for anemia, including hematocrit, hemoglobin, erythrocyte protoporphyrin • Tests for iron status, including serum iron, serum ferritin, total iron binding concentration, and percent saturation • Tests for malabsorption • Tests for specific nutrient deficiencies	Nutrition intervention may be indicated by abnormal lab test results.
Clinical/Medical History Complete a health history by interviewing caregiver(s) and reviewing medical records. Pay special attention to nutrition risk factors identified in nutrition screening. Include the following in data collection: • Medical diagnosis • Frequency of infections • Reflux/vomiting not able to be managed with routine precautions (medications, positioning, etc.) • Elimination patterns • Maturation stage, age of onset of puberty • Possible medication-nutrient interactions • Family history of diseases • Family growth history • Dental health	Nutrition intervention indicated if any unresolved nutrition concern. Refer to primary care provider for follow-up and referral to appropriate medical and pediatric feeding specialist(s).
• Elimination patterns[6] • Maturation stage, age of onset of puberty • Possible medication-nutrient interactions[7] • Family history of diseases • Family growth history • Dental health	
Dietary	
Assess dietary intake with a 3 to 7-day food record and diet history. When requesting a food record, provide both oral and written instructions. When interviewing for a diet history, include both the child and caregiver(s) if possible. Obtain the following data: • Type, brand name, and amount of food, beverage, or formula eaten or tube-fed • Preparation method for cooking foods and for mixing formula • Time of each meal, snack, or feeding • Cooking facilities available • Location of feedings (e.g., day care center, school, home, restaurant) • Dietary supplements (e.g., vitamins, minerals, energy dense liquids) • Complementary and alternative therapies/supplements • Intake of non-food items (pica) Adequacy of food intake may be determined by comparison with age-appropriate food group plan or by computer analysis and comparison with DRIs for age and sex. Consider: • Level of physical activity or ambulation. • Cultural and familial food practices. • Pertinent historical data related to feeding: breastfeeding, amount of formula milk used, age of introduction of solid foods, variety of solids provided. • Influences on the validity of food record (e.g., illnesses, meals eaten away from home, losses from reflux).	For nutrient recommendations for specific conditions and disorders, refer to appropriate section in this manual.

[6] See Chapters 6 and 7.
[7] See Chapter 5.

continued...

Table 1-2: Nutrition Assessment (continued)	
Assessment Activities	**Example Indicators for Nutrition Intervention**
Feeding Skills[8] and Behavior[9] Complete a feeding history by interviewing caregiver(s) and reviewing health, therapy, and assessment records. Observe child while eating or being fed. Consider the following factors: • Positioning of child • Appropriateness of feeding environment • Oral-motor development and coordination • Self-feeding skills • Behavior problems related to feeding • Child-caregiver interactions during feeding[10]	Multi-disciplinary intervention with pediatric feeding specialists such as speech language pathologist/therapist, occupational therapist, registered dietitian nutritionist (RDN), public health nurse, behaviorist, and/or social worker indicated if any of the following: • Delayed or abnormal feeding skills • Neurological or oral-motor problems • Behavior problems interfering with feeding • Suboptimal scores on feeding assessment tools

[8] See Chapter 8.
[9] See Chapter 9.
[10] Assessment tools for documenting inappropriate or at-risk child-caregiver interactions during feeding are the NCAST Feeding Scale (up to age 1) and the CHATOOR Feeding Scale (up to age 3) (9, 10).

Table 1-3: Pediatric Malnutrition (11)

Primary Indicator	Mild Malnutrition	Moderate Malnutrition	Severe Malnutrition
Weight-for-height z-score*	−1 to −1.9 z-score	−2 to −2.9 z-score	−3 or greater z-score
BMI-for-age z-score*	−1 to −1.9 z-score	−2 to −2.9 z-score	−3 or greater z-score
Length/height-for-age z-score*	No data	No data	−3 z-score
Mid–upper arm circumference*	Greater than or equal to −1 to −1.9 z- score	Greater than or equal to −2 to −2.9 z-score	Greater than or equal to −3 z-score
BMI-for-age z-score*	−1 to −1.9 z-score	−2 to −2.9 z-score	−3 or greater z-score
Weight gain velocity (<2 years of age)	Less than 75%a of the normb for expected weight gain	Less than 50%a of the normb for expected weight gain	Less than 25%a of the normb for expected weight gain
Weight loss (2–20 years of age)	5% usual body weight	7.5% usual body weight	10% usual body weight
Deceleration in weight for length/height z-score	Decline of 1 z-score	Decline of 2 z-score	Decline of 3 z-score
Inadequate nutrient intake	51%−75% estimated energy/protein need	26%−50% estimated energy/protein need	≤25% estimated energy/ protein need

* May be used when only one data point is available

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CHAPTER 2

ANTHROPOMETRICS

Karen Murphy, MS, RD, CD
Updated by Sharon Feucht, MA, RD, CD
Updated 2021 by Sarah Harsh, MS, RDN, CD

The term anthropometry refers to comparative measurements of the human body. The primary measures used as indices of growth and development include stature (length or height), weight, and head circumference (for young children under 2 years of age). The secondary measures used to estimate body composition include triceps skinfold thickness, subscapular skinfold thickness, and mid-upper arm circumference. Growth is an important index of a child's nutritional status and should be monitored on a regular basis.

Stature (length or height), weight, and head circumference are typically evaluated by comparing individual measurements to population data, represented by percentile curves on a growth chart. The CDC currently recommends using the 2006 World Health Organization (WHO) growth charts for children aged 0-24 months and the CDC growth charts for children 2-20 years (1,2). These charts were developed by collecting measurements from healthy children and are intended as clinical tools to assess the nutritional status and general health of infants, children, and adolescents. A single measurement is not enough information to identify a growth concern. However, for the typically developing child, height- or weight-for-age either below the 5th percentile or above the 95th percentile is a reason for further evaluation. To assess and monitor the nutritional status of a child or youth with special health care needs who does not necessarily meet the criteria of the growth charts, it is necessary to carefully evaluate a pattern of measurements obtained at regular intervals.

Body mass index (BMI) or weight-for-length are indirect measures of body composition, and comparing individual measurements to percentiles on a growth chart or z-scores (a measure of the number of standard deviations from the mean) can be used to identify atypical growth patterns in typically developing children. Current recommendations identify malnutrition/undernutrition as a BMI z-score of <-1 (3), overweight as a BMI between the 85-94th percentile and obesity as a BMI >95th percentile (4). For the child or youth with special health care needs, these parameters may not be reliable indicators of atypical growth. However, they are useful in screening for children and youth who are at risk for growth problems. For some children and youth with special health care needs, poor growth or excessive weight gain must be confirmed with longitudinal measurements and in many cases, additional anthropometric parameters to estimate body composition. Furthermore, the growth patterns characteristic of the particular disease or disorder and the child's growth history must be considered.

For anthropometric parameters to be valid indices of growth status, they must be highly accurate. This requires precise measurement techniques. Appropriate use of growth charts requires that measurements be made in the same manner in which the reference data were secured (5,6). In order to measure a child accurately, the individual performing the measurement must be properly trained, and reliable equipment must be available. For some children and youth with special health care needs, it can be challenging to make accurate measurements because of factors such as contractures and low muscle tone.

This chapter provides information on recommended equipment and measuring techniques including special considerations for obtaining measurements from the child or youth with special health care needs. Guidelines for measuring and interpreting length, crown-rump length, height, sitting height, arm span, weight, BMI, head circumference, triceps skinfold, mid-upper arm circumference, and subscapular skinfold are also provided. The concepts of growth as an index of nutritional status and ideal body weight (IBW) are also discussed. Measurement techniques, equipment required, advantages, and limitations are summarized in Table 2-1.

Primary Measures

Head Circumference (5,6)

Head circumference is an important screening tool in infants and young children because it is closely related to brain growth. A rapid increase in the rate of growth may indicate hydrocephalus. A decrease in the rate of head growth may indicate a developmental delay. Decreases in the rate of head growth have been seen in children who are severely undernourished. Children with slow head growth frequently have poor linear growth as well. Head circumference should be measured routinely until at least 24 months of age. Parental head circumferences of infants whose head circumferences are atypical should also be measured, as head circumferences of parents and their offspring are typically closely associated. The proper technique for measuring head circumference is shown in Figure 1.

Figure 1. Measuring Head Circumference

Equipment and technique for measurement of head circumference

1. Use a flexible, non stretchable measuring tape.
2. Position the child standing or in a sitting position in the lap of their caregiver. If possible, remove any barrettes or hair styles that may make the measurement less accurate.
3. Place the lower edge of the measuring tape just above the child's eyebrows, above the ears, and around the occipital prominence at the back of the child's head.
4. Pull the tape snugly to compress the hair. The objective is to measure the maximal head

circumference.
5. Repeat the measurement twice or until two measurements agree to 0.1 cm or 1/16 in.
6. Record the numeric value and plot it on the appropriate growth chart.
7. If the measurement appears larger or smaller than expected when plotted, check the accuracy of plotting and recheck the measurement. If there is a rapid increase in the child's head circumference, they should be seen by a physician.

Guidelines for interpretation of head circumference

The WHO percentiles (age 0-24 months) for head circumference are the most readily available, as they are printed with the WHO percentiles for length, weight, and weight for length. For many children, it is useful to continue following head circumference measurements past the age of 2 years. For these children, there are two growth charts available with percentiles from 0 to 18 years. The Nellhaus charts are widely available and were developed from international, interracial data (7). The more recent charts from Rollins et al. were developed from several different US data sources and have the advantage of including more detailed percentile intervals (8). Premature infant growth charts include percentiles for head circumference as well as for length and weight. The most commonly used preterm growth charts include Fenton (updated 2013) and Olsen (2010) (See Chapter 14). When monitoring head growth, it is important to consistently plot measurements on the same chart and to look for consistent patterns in head growth over time.

Stature (Length and Height)

Stature is measured in two ways: recumbent length for the child younger than 24 months of age and standing height for children and youth older than 24 months (5,6). Alternative measurements (e.g. crown-rump length, sitting height, and arm span) can also provide information about a child's stature.

Contractures about the hips, knees, and ankles can interfere with an accurate stature measurement. Crown-rump length or sitting height measurements are often useful estimates of stature for children and youth with contractures of the lower body. These measurements will not correlate directly with height or length, but can indicate a child's rate of growth when plotted on CDC growth charts. Although the measurements will be below the 3rd percentile for age, they will show whether or not the child is following a consistent growth curve. The stature of children and youth with involvement of the lower body only (e.g. some children with myelomeningocele) can be estimated by using arm-span measurements. However, for children with contractures of the upper extremities such as in cerebral palsy, accurate arm span measurements are also difficult. For those children and youth who have contractures of the arm, tibia length, though less accurate, is sometimes used with a formula to estimate stature (9).

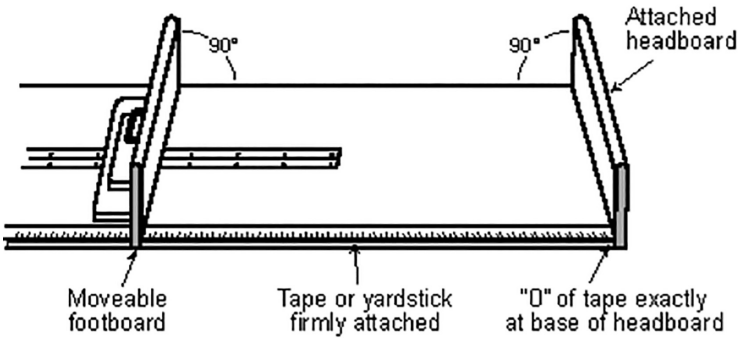
Length

For children who are younger than 24 months of age, measure recumbent length. Older children and youth who are unable to stand may also be measured in the recumbent position; however, it should be noted on the growth chart that the measurement is length, not height.

Equipment for length measurement

In order to have accurate recumbent length measurements, it is important to have a good quality length-measuring device. The infant length board should have a fixed headboard and a movable footboard that are perpendicular to the surface on which the child is lying. A measuring tape, marked in millimeters or 1/8 inch segments, is needed along one or both sides of the table, with the zero end at the end of the headboard (5,6). The required features of an infant length board are shown in Figure 2. The proper technique for measuring length is shown in Figure 3.

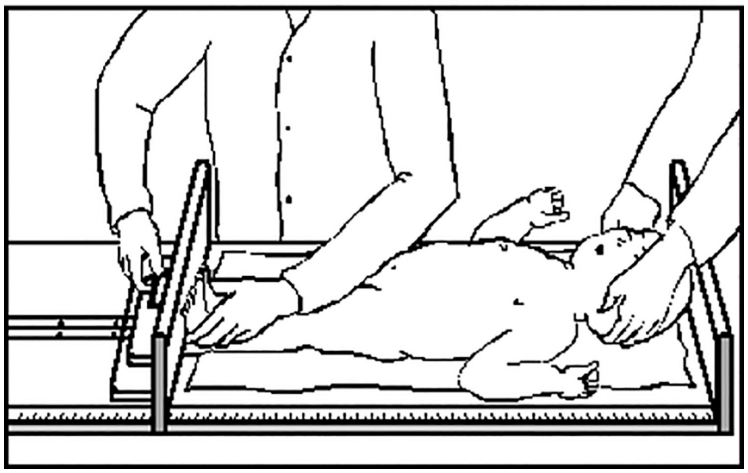
Figure 2. Infant Length Board



Technique for length measurement (5,6)

Clothing that might interfere with an accurate measurement, including diapers, should be removed. Two people are required to measure length accurately as shown in Figure 3.

Figure 3. Measuring Infant Length



Person A

1. Hold the child's head with the crown against the headboard so that the child is looking straight upward.
2. Make sure that the trunk and pelvis are aligned with the measuring device.

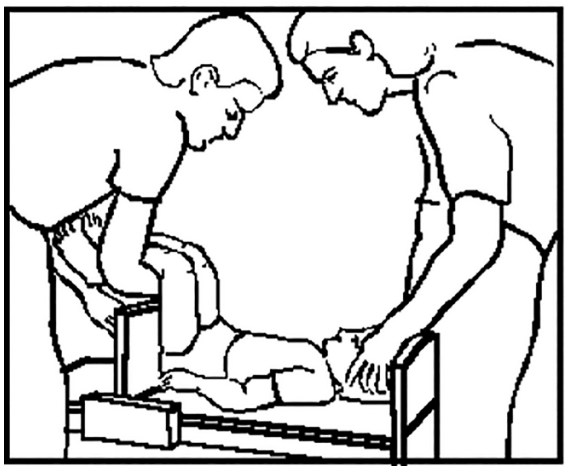
Person B

3. Straighten the legs, holding the ankles together with the toes pointed directly upward.
4. Move the footboard firmly against the soles of the child's feet.
5. Read the measurement to the nearest 0.1 cm or ⅛ in.
6. Repeat the measurement until two measurements agree within 0.2 cm or 1/8 inch.
7. Record the numeric value and plot length for age on the 0 to 24 month growth chart appropriate for age and sex assigned at birth.

Technique for length estimation: crown-rump length

Use the same equipment and technique as that described for measuring length, except bend the child's legs at a 90-degree angle and bring the footboard up against the buttocks. The proper technique for measuring crown rump length is shown in Figure 4.

Figure 4. Measuring Crown-Rump Length



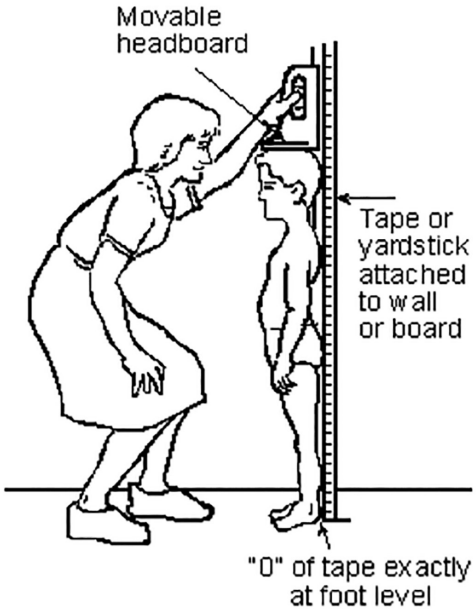
Height

It is important to plot standing height measurements on the growth charts for 2 to 20-year olds, because the percentiles are adjusted for the difference between recumbent length and standing height. If a child or youth is unable to stand, a length measurement may be used, but it should be noted as a length measurement.

Equipment for height measurement

Use a measuring board with an attached, movable headboard (stadiometer). If this is not available, use a non-stretchable tape measure attached to a vertical, flat surface, like a wall or a door jam with no baseboard, and equipment that will provide an accurate right angle to actually take the measurement. The movable measuring rod that is attached to a platform scale is too unsteady to ensure accurate measurements. The features of an accurate stadiometer are shown in Figure 5.

Figure 5. Stadiometer



Technique for height measurement (5,6)

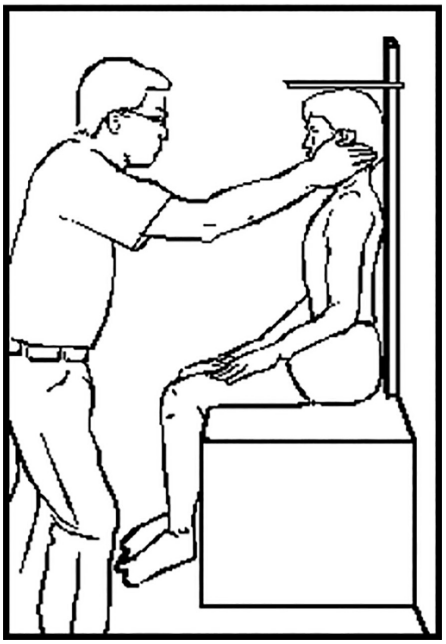
Two people may be required for accurate measures of younger children; however, usually only one measurer is required for most older children.

1. Measure the child with underclothes only, if possible, or with non bulky clothing and no shoes.
2. Have the child stand with heels together and touching the floor, knees straight, arms at sides, shoulders relaxed, and shoulder blades, buttocks, and heels touching the wall or measuring surface.
3. Have the child look straight ahead with their line of vision perpendicular to the body.
4. Lower the headboard or right angle onto the crown of the child's head.
5. Read the measurement to the nearest 0.1 cm or 1/8 inch. When reading, make sure your eyes are level with the headboard.
6. Repeat the measurement until two measurements agree within 0.1 cm or 1/8 inch.
7. Record the numeric value and plot height for age on the appropriate growth chart.

Technique for stature estimation: sitting height

Use the same equipment as that described for measuring standing height, except have the child or youth sit on a box of known height and subtract the height of the box from the measurement obtained. The box should be high enough so that the child's legs hang freely. Sitting height should not be measured with the child sitting on the floor or on a box with legs extended outward in a 90-degree angle (5). The proper techniques for measuring sitting height are shown in Figure 6.

Figure 6. Measuring Sitting Height

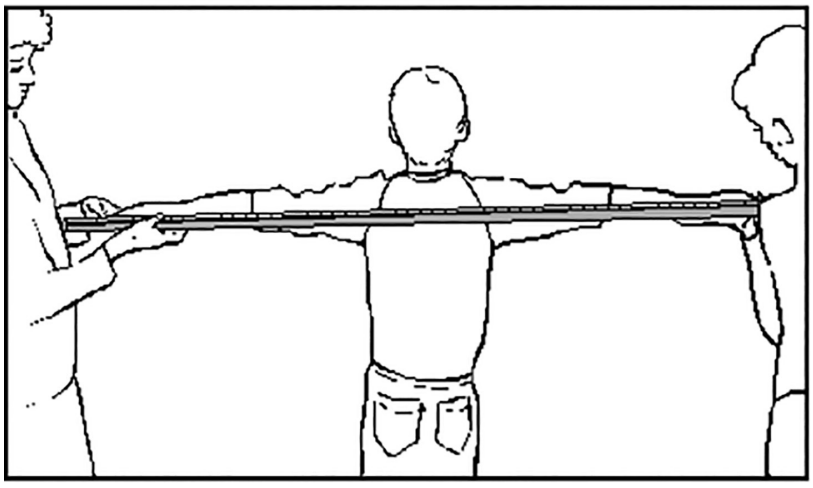


Technique for stature estimation: arm span (9)

Arm span is defined as the greatest distance between the tips of the extended middle fingers of the right and left hands when the arms are fully extended to the sides at right angles to the body and the back is straight. Measurement of the arm span is useful for estimating the stature of persons with lower extremity contractures or lower body paralysis. For the typically developing child over age six, the ratio of arm span to height has been found to be 1:1. This may not be the case for the child or youth with special health care needs; however, monitoring an individual's arm span measurements over time can provide some information about growth.

Arm span is not an adequate substitute for stature in persons with contractures of the upper extremities (e.g., in spastic quadriplegia) because these individuals cannot fully extend their arms and fingers. Also, arm span cannot accurately estimate stature in young children (younger than 5 to 6 years) because the proportions of limb length and trunk length to total body length are different for younger children compared to older children and youth.

Figure 7. Measuring Arm Span with an Anthropometer



Equipment for arm span measurement

Arm span measurements are made with an anthropometer, a stainless steel detachable rod approximately seven feet long with etched gradations to 0.1 cm or 1/8 inch and one movable sleeve (9). The proper technique for measuring arm span is shown in Figure 7.

Technique for arm span measurement

Two persons are needed to measure arm span.

Person A

1. Have the child sit in an erect position with arms outstretched.
2. Hold the fixed end of the anthropometer at the tip of the middle finger of one of the child's hands.

Person B

3. Position the sleeve at the tip of the middle finger of the child's other hand with the anthropometer going across the child's back.
4. Have the child stretch their arms while the movable sleeve is adjusted to the maximum arm span.
5. Repeat the measurements until two measurements agree within 0.1 cm or 1/8 inch.
6. Record the actual numeric value, and plot as height for age on the appropriate growth chart. On the chart, note that arm span was the technique used to estimate stature.

Weight

Infants and toddlers can be accurately weighed on an infant scale up to the weight limit stated by the manufacturer. An accurate measure of weight is critical—it is more valuable to obtain an accurate bi-annual weight than a series of inaccurate monthly weights.

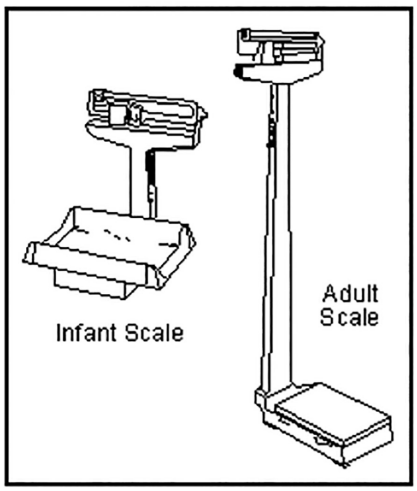
Equipment for weight measurement

Use a calibrated beam balance scale with non-detachable weights or a digital scale with a "strain-gauge" mechanism. To weigh infants and young children who cannot stand, use a pan-type or bucket seat-type pediatric scale that is accurate to within 0.01 kg or ¼ oz. For older children and youth who can stand, use a platform beam scale or an electronic scale that is accurate to within 0.1 kg or ¼ lb. Do not use a spring-type bathroom scale which, with repeated use, will not maintain the necessary degree of accuracy. For children who are too large for the infant scale but cannot stand, use a platform scale on which a wheelchair can be placed, or a bed scale. Since this type of specialty scale is not available in many communities, it can be difficult to regularly monitor the weight of children and youth with special health care needs. An alternative is to weigh the child's caregiver holding the child, weigh the caregiver alone, and subtract the caregiver's weight from the weight of both individuals. If this method is used, it is important to note this on the growth chart. If the child can sit independently but is not able to stand, use a chair scale.

Frequently check and adjust the zero weight on the beam scale by placing the main and fractional sliding weights at their respective zeros and moving the zeroing weight until the beam balances at zero. If a pad or diaper is used to make the pan more comfortable, place it in the pan before the zero adjustment is made; otherwise, the weight of the pad or diaper must be subtracted from the weight of the child each time a measurement is made. At least two or three times per year, have the accuracy of the scale checked with a set of standard weights by a local dealer or an inspector of weights and measures. Equipment for measuring weights is shown in Figure 8.

Technique for weight measurement (5,6)

Figure 8. Scales



Infants

1. Remove the infant's clothing and diaper.
2. Center the infant in the scale tray.
3. Weigh infant to the nearest 0.01 kg or ¼ oz.
4. Repeat the measurement until two measurements agree to within 0.02 kg. or ½ oz.
5. Record the numeric value and plot weight for age and weight for length on the appropriate growth chart(s).
6. Record any information about conditions that might have interfered with an accurate weight measure (e.g., infant was moving).

Children and Youth (able to stand)

1. Weigh the child with only lightweight undergarments or a hospital gown and no shoes.
2. Have the child stand in the center of the scale's platform touching nothing and with heels together.
3. When the child is standing still, read the scale to the nearest 0.1 kg or ¼ lb.
4. Repeat the measurement until two measurements agree to within 0.2 kg or ½ lb.
5. Record the numeric value and plot weight for age and weight for height on the appropriate growth chart(s).
6. Record any information about conditions that might have interfered with an accurate weight measure (e.g., child was moving).

Guidelines for Interpretation of Length, Height, and Weight (1,2)

Various growth charts have been developed from reference data for length, height, and weight. The CDC currently recommends using the 2006 WHO growth charts for children aged 0-24 months and the CDC growth charts for children 2-20 years. The WHO charts are separate for girls and boys based on sex assigned at birth and include weight for age, length for age, weight for length, and head circumference. The CDC growth charts for girls and boys (based on sex assigned at birth) ages 2 years to 20 years include weight for age, height for age, weight for height (for children 77-121 cm only), and BMI for age. Any growth chart is most useful if measurements are accurately obtained and plotted on a regular basis so that the child's growth pattern can be observed. Information about these growth charts and downloadable versions of the charts can be found on the CDC website: http://www.cdc.gov/growthcharts/.

On the CDC 2000 charts, Body Mass Index (BMI) replaces the weight for stature curves for children and youth over age 2 years. A separate weight for height chart is available for children 77-121 cm tall (approximately 2-5 years of age). Charts that include the 3rd and 97th percentiles for weight and stature for age are also available. An 85th percentile line has been added to the BMI for age

chart to aid in assessing risk of overweight.

The WHO charts for infants aged 0-24 months were constructed from longitudinal data from breastfed infants at 6 sites around the world, including the US, who lived in conditions that were most likely to support optimal growth. These charts were developed to represent the growth of infants under ideal conditions. The CDC charts for 2-20 year olds are based on cross-sectional data from the five previous NHANES studies, and represent the racial diversity of the US. NHANES III data was not used for weight and BMI for age percentiles for children over age 6 years because of the trend toward obesity in this age group. As a result of these differences in methodology, the WHO charts describe how children should grow (prescriptive) regardless of time and place, while the CDC charts describe how children and youth do grow (descriptive) in a particular time and place. The WHO has also produced growth charts for children aged 2-5 years. The methods made to create these charts are similar to the methods used to create the CDC charts, and the use of the CDC charts for this age range is recommended.

Body Mass Index (BMI) (2, 10)

Body mass index (BMI) is a calculation that is used as an indirect measure of body composition in children over 2 years of age. It has been recommended as a non-invasive and clinically convenient measure. It can be used to screen for over- and undernutrition. BMI is expressed as a ratio of weight in kilograms to height in meters squared:

$$\text{BMI} = \frac{\text{weight in kilograms}}{(\text{height in meters})^2}$$

OR

BMI = (weight in kilograms) ÷ (height in meters) ÷ (height in meters)

BMI can also be calculated using English units (8):

BMI = [Weight (pounds) ÷ Height (inches) ÷ Height (inches)] x 703

The calculated BMI adds a useful dimension to the assessment of body composition if accurate stature (length or height) and weight measurements are obtained. This index of weight relative to length or stature can be used to monitor changes over time. With this addition, clinicians can compare a child's BMI to the BMI of their peers. Because growth parameters change, no single BMI is ideal during childhood and adolescence.

Weight Gain Velocity

Comparing weight gain velocity (the amount of weight gain over a defined period of time) to mean rates can be helpful to identify deviations from growth charts that would be difficult to identify on weight and length for age charts. For example, a child weighing 7 kg at 12 months of age and 8.8 kg at 18 months plots below the 5th percentile for weight for age on the WHO charts, but shows a weight gain velocity greater than the 85th percentile. This child, although below the 5th percentile

for weight for age, is demonstrating a rate of weight gain that is faster than the mean. Weight gain velocity percentiles from the WHO charts are available on the WHO website (11).

Specialty Growth Charts

Growth charts exist for children and youth with several different specific conditions. These charts should be used as an additional tool for interpretation of growth after data have been plotted on the WHO or CDC charts. Many of these specialty growth charts are based on cross sectional data from small groups of children with specific disorders and do not necessarily reflect ideal rates of growth. The user of the specialty growth chart should also consider when the data for the growth chart was gathered and if treatments or outcomes for the condition have changed since that time. As with all growth charts, the trend in measurements over time is more important than the percentile of any one measurement.

Growth charts for premature infants that attempt to reflect intrauterine growth rates have been produced by several different researchers; each set has benefits and drawbacks. However, instead of premature infant charts, many practitioners use the WHO growth charts and correct for the child's prematurity. It is important to document that measurements of age are corrected for prematurity. It is best to continue to correct for prematurity until the child's growth is plotted on the charts for 2-20 year olds.

"Ideal" Body Weight

Ideal body weights were previously used to assess nutrition status. However, because children grow at different rates, it is impossible to determine an absolute "ideal" weight based solely on age or height. Nutrition status is better understood looking at changes in weight, stature, and weight-for-stature or BMI Z-scores over time (3).

Malnutrition (Undernutrition)

Malnutrition (undernutrition) is the current term used to refer to children and youth who are not meeting expected growth goals. These children may have previously been described as having failure to thrive. The consensus statement of the AND and ASPEN suggest that malnutrition more accurately reflects the etiology of the problem instead of the symptom. It is well known that children and youth with special health care needs are more at risk for both acute and chronic malnutrition. Mild malnutrition has been defined as a BMI or weight for length z-score of <-1 to -2, moderate malnutrition as a BMI or weight for length z-score of <-2 to -3, and severe malnutrition as a BMI or weight for length z-score of <-3 (3). See Chapter 1, Table 1-3 for more information.

Secondary Measures

Triceps Skinfold and Mid-upper Arm Circumference (4,5)

Together, triceps skinfold thickness and mid-upper arm circumference are used to calculate arm muscle circumference, arm muscle area, and arm fat area, which are indicators of body fat and muscle stores when compared to population percentiles (5,9). For typically developing children, the calculations of arm muscle circumference, arm muscle area, and arm fat area provide a better estimate of body composition than triceps skinfold alone.

Accurate measurements of triceps skinfold thickness and mid-upper arm circumference are difficult to obtain by an inexperienced or untrained measurer. Measurement error is likely to be higher when measuring young children because it is difficult to maintain the child in the proper position while the measurement is being performed. It is also difficult to separate fat from muscle tissue (9). These secondary measurements are useful only if obtained with precise and accurate technique that is developed with training and practice. In the course of training, the measurements must be validated by a person experienced with skinfold thickness techniques. Only calibrated calipers should be used for measuring skinfold thickness; plastic calipers are not accurate. In order to minimize error and variation in technique, serial measurement should be taken by the same clinician whenever possible. For further information on these measurements see Frisancho (12), Guiney (13), and Tanner (14).

Recommended guidelines for interpretation have been published, and an individual's measurements can be compared to population reference data (12). These interpretations, however, are based on assumptions of the bone diameter and the distribution of muscle and fat around the bone of typically developing persons; these assumptions may be inaccurate for persons with physical differences. The best use of these measurements for children and youth with special health care needs is for assessing changes over time (e.g., increases in fat and muscle stores in the undernourished child and decreases in fat stores in the overweight child). Skinfold measurements are not appropriate for children or youth with subcutaneous edema.

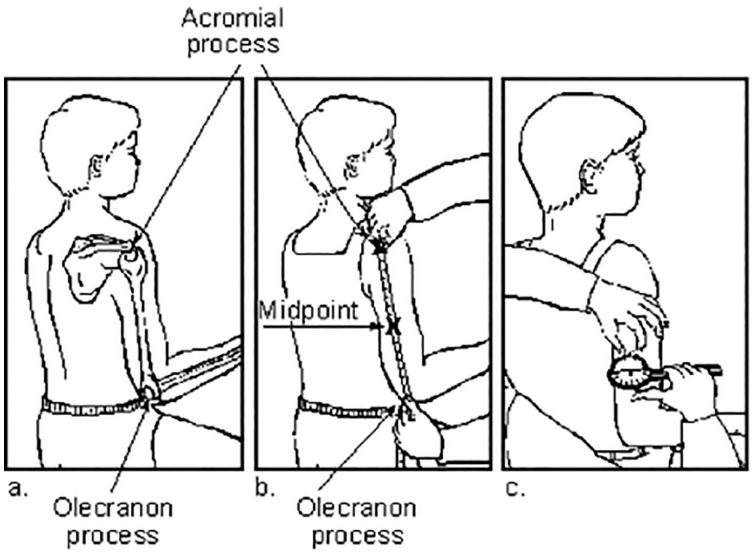
Equipment and technique for triceps skinfold measurements (5, 9,13)

1. Use an accurate skinfold caliper, such as the Lange or the Holtain, and a flexible, non-stretchable tape measure.
2. On the child's right side, find the acromion process and the olecranon process (tip of elbow). These processes are shown in Figure 9.
3. Using a tape measure, find the midpoint between the acromion process and the olecranon process and mark it with a pen as shown in Figure 9.
4. Position the child with their right arm completely relaxed and hanging by their side.
5. Pick up the skinfold overlying the triceps muscle, 1 cm above the midpoint mark.
6. At the midpoint mark, apply the jaws of the caliper to the skinfold while continuing to hold

the skinfold above the mark, as shown in Figure 9.

7. Permit the jaws of the caliper to exert full strength as the trigger lever is released, without "snapping" it.
8. Read the dial to the nearest 0.5 mm.
9. To minimize error, repeat the measurement 3 times and average the values. Make sure that there is no tissue compression with the repeated measurements.
10. Record the numeric value and compare it to reference data and/or previous measurements.

Figure 9. (a) Locating the acromion and the olecranon processes, (b) measuring the midpoint, and (c) measuring triceps skinfold thickness



Equipment and technique for mid-upper arm circumference (5,9)

1. Use a flexible, non-stretchable tape measure.
2. Position the child with their right arm completely relaxed and hanging by their side.
3. Measure the circumference of the right arm at the midpoint mark (midway between the acromial and olecranon processes as shown in Figure 9).
4. Wrap the tape around the arm so that it is touching the skin but not perpendicular to the long axis of the arm.
5. Measure to the nearest 0.1 cm.
6. Repeat the measurement until two measurements agree within 0.2 cm.
7. Record the numeric value and compare it to reference data and/or previous measurements.

Calculating arm muscle circumference, arm muscle area, and arm fat area (5,12)

The mid-upper arm circumference (C) is converted to mm (c) and used with triceps skinfold thickness (T) to calculate upper arm area (A), upper arm muscle area (M), and upper arm fat area (F). Equations for these calculations are provided below.

- Upper arm area (mm^2) = $\left[\frac{\pi}{4}\right] \times \left[\frac{c}{\pi}\right]^2 = A$
- Upper arm muscle area (mm^2) = $\frac{(c-\pi T)^2}{4\pi} = M$
- Upper arm fat area (mm2) = F = A – M

Guidelines for interpretation of upper arm indices of fat and muscle stores

Percentiles for triceps skinfold, mid-upper arm circumference, arm muscle circumference, arm muscle area, and arm fat area for White males and females 1 to 75 years have been published by Frisancho (12). These indices are appropriate for assessing an individual's fat and muscle stores, but it is important to remember that these are reference data for typically developing White persons.

Subscapular Skinfold

Subscapular skinfold thickness is a useful measurement for estimating fat stores, especially when used in conjunction with triceps skinfold thickness and mid-upper arm circumference. Percentiles for subscapular skinfold thickness for typically developing males and females ages 0 to 19 years have been developed by Tanner and Whitehouse (14). It has been suggested that the best use of the subscapular skinfold measurement in managing children and youth who are overweight or underweight is to evaluate individual change over time.

Equipment and technique for subscapular skinfold (9)

1. Use an accurate skinfold caliper, such as the Lange or the Holtain.
2. Pick up the subscapular skinfold just under the shoulder blade, following the natural fold of the skin.
3. With a pen, mark the midpoint of the fold.
4. Holding the skinfold approximately 1 cm from the midpoint mark, apply the jaws of the caliper to the skinfold so that the mark is midway between the jaws, as shown in Figure 10.
5. Permit the jaws of the caliper to exert full strength as you release the trigger lever, but do not allow them to "snap" and pinch the child.
6. Take the reading right after the first rapid fall. Read to the nearest 0.1 cm.
7. Repeat the measurement three times and recor

Figure 10. Measuring Subscapular Skinfold Thickness

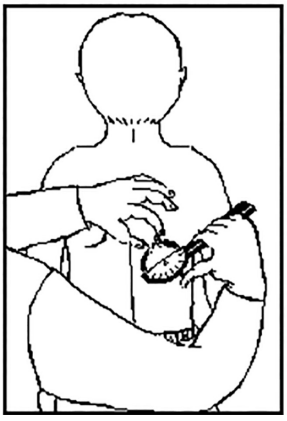


Table 2-1: Measurement Techniques

Technique	Equipment	Advantages	Limitations	Tolerance levels[1]
Stature				
Length	Length board	Direct measure of stature	Hard to do if contractures	0.1 cm
Height	Stadiometer	Direct measure of stature	Hard to do if contractures	0.1 cm
Stature Estimation				
Crown-rump	Length board	Provides estimate of stature	Limited data available	0.1 cm
Sitting height	Stadiometer, sitting box	Provides estimate of stature	Must be able to sit independently	0.1 cm
Arm span	Anthropometer	Provides best estimate of stature; 1:1	Requires full arm extension	0.2 cm
Upper arm length	Anthropometer	Provides estimate of stature	Difficult to interpret	
Knee height	Anthropometer	Provides estimate of stature	Useful if contractures, difficult to interpret	
Tibia length	Anthropometer	Provides estimate of stature	Not used if <2 years old, difficult to interpret	
Weight	Calibrated Scale			0.1 kg (infants: 20 g or 0.04 kg)
Skinfolds				
Subscapular	Caliper	Provides estimate of total body fat	Difficult to maintain technique; inappropriate for obesity; not for <1 year	3 mm
Triceps	Caliper, flexible tape	Provides estimate of percent body fat	Difficult to maintain technique; inappropriate for obesity; not <1 year	3 mm
Circumferences				
Head	Flexible tape	Direct measure of head circumference	None	0.1 cm
Mid-arm	Flexible tape	Estimator of total body fat	May be difficult to interpret	0.2 cm

[1] The measure should be reproduced with a difference no greater than the value in this column.

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MCHB (Maternal and Child Health Bureau) Growth Charts Training – Self-study material, including modules about equipment, measurement technique, and using the CDC Growth Charts with children with special health care needs. http://depts.washington.edu/growth/index.htm

CHAPTER 3

PHYSICAL ACTIVITY FOR CHILDREN AND YOUTH WITH SPECIAL HEALTH CARE NEEDS

Kathleen Washington, PT, PhD and Kay Kopp, OTR/L
Updated by Kay Kopp, OTR/L

Introduction

A 2017-18 study completed through the Child and Adolescent Health Measurement Initiative showed that 18.5 percent of U.S. children under the age of 18 had a special health care need. This figure represents 13.5 million children (1).

These children are at greater risk than peers without disabilities to develop poor health, including obesity, cardiovascular disease, and musculoskeletal limitations because they are not appropriately physically active and fit.

The most frequently identified barriers to participation of these children in sports and physical activities are the child's functional limitations (18%), high costs (15%), and lack of nearby facilities or programs (10%) (2). The child's perceived self-competence, time, social support from schools and communities, and family and child preferences are additional factors influencing participation. Families who engage in physical activities are more likely to encourage similar participation for their children with special health care needs. Interestingly, environmental and family factors seem to be more significant determinants of participation than characteristics of the children themselves (2).

The primary goals for increasing physical activity in these children are to reverse deconditioning secondary to impaired mobility, optimize performance of functional activities, and enhance well-being. Regular physical activity is essential for the maintenance of normal muscle strength, flexibility, and joint structure and function, and may prevent, minimize, or slow the loss of function often associated with disabling conditions. Other benefits of physical activity include improvements in confidence and self-esteem (3).

Definition and Types of Physical Activity

The term "physical activity" describes many forms of movement that involve the large skeletal

muscles and require significant energy expenditure. Physical activity is defined by its duration (amount of time), intensity (rate of energy expenditure), and frequency (number of sessions per time period). As described in *Bright Futures in Practice: Physical Activity* (4) there are several types of physical activity:

1. Aerobic: Light to vigorous intensity physical activity that requires more oxygen than sedentary behavior and thus promotes cardiovascular fitness and other health benefits (e.g. jumping rope, playing soccer or basketball)

2. Anaerobic: Intense physical activity that is short in duration and requires a breakdown of energy sources in the absence of sufficient oxygen. Energy sources are replenished as an individual recovers from the activity. Anaerobic activity (e.g. sprinting during running or biking) requires maximal performance during a brief period.

3. Lifestyle: Physical activity typically performed on a routine basis (e.g. walking, climbing stairs, raking the yard) which is usually light to moderate in intensity.

4. Physical activity play: Play activity that requires substantial energy expenditure (e.g. playing tag, jumping rope)

5. Sports: Physical activity that involves competition, scorekeeping, rules, and an outcome that is not known in advance. Sports can be divided into categories such as individual (e.g. gymnastics, swimming) dual (e.g. tennis) and team (e.g. basketball)

Most of the above are weight-bearing activities, which contribute to the growth of healthy bones in children and adolescents, and provide numerous other health benefits. Many children and adolescents choose not to participate in competitive team sports but can still gain health benefits by participating in individual activities such as biking or yoga. One of the most important criteria for promoting physical activity for children and adolescents is that it is enjoyable for them.

Two broader categories of physical activity for children and adolescents are lifestyle and structured (4). Examples of lifestyle physical activities are walking, playing, and doing chores. Structured physical activities consist of 1) physical education (PE) programs at school and 2) extracurricular activities (e.g. baseball team), which can occur in either in school or non-school settings. Structured physical activities generally provide more intense physical activity than lifestyle activities, and are more likely to help maintain weight and result in improved strength and cardiovascular fitness.

The Importance of Physical Activity in Health Promotion

The role of physical activity in the prevention of disease and health promotion for all children and adults has been advocated by many organizations.

In 2018, the U.S. Department of Health and Human Services published Physical Activity Guidelines for Americans 2nd Edition (5). The guidelines for preschool children (ages 3 through 5 years) recommend physical activity throughout the day to enhance growth and development. Those 6 through 17 years should engage in 60 minutes or more of moderate-to-vigorous daily physical activity to include aerobic activity, muscle strengthening, and bone strengthening. (5)

Children with disabilities are urged to meet these guidelines when possible and avoid being inactive. Other organizations, such as the American College of Sports Medicine (6) and the National Center on Physical Activity and Disability (7) have promoted the health benefits of physical activity, and articulated the public health concerns that inactivity presents. As children and you with special health care needs are at serious risk for additional health consequences such as obesity and musculoskeletal impairments as a result of a more sedentary lifestyle, increasing their levels of physical activity should be a life-long objective.

The International Classification of Functioning, Disability and Health, known more commonly as ICF, is the framework developed by the World Health Organization (WHO) for measuring health and disability at both individual and population levels (8). The ICF is a classification of three domains of human functioning: *body structures and functions*, *activities*, and *participation*. (See Fig. 1) *Body functions* refers to the physiological function of body systems, while body structures refers to anatomical body parts, such as organs and muscles. *Activities* refers to a person's performance of tasks such as getting dressed, walking, or running. Participation refers to the nature and extent of a person's involvement in daily life situations, such as attending school, socializing, or playing sports (recreation). A primary goal of health care professionals is to assist children with special health care needs in participating as fully as possible in the life of their family and community. Since an individual's functioning and disability occurs in a context, the ICF also includes *personal factors* (e.g. personality, age, lifestyle) and *environmental factors* (e.g. physical, economic, social). These contextual variables allow a more holistic view of disability on the person's functioning.

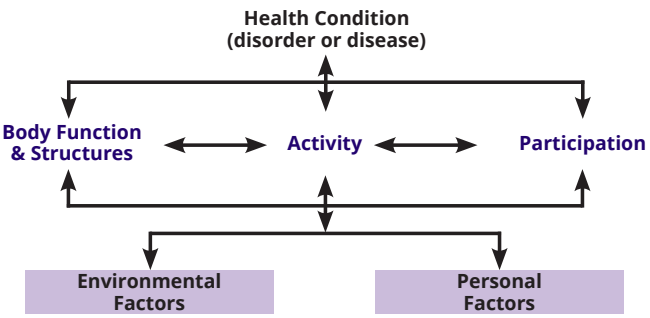

Fig. 1: The International Classification of Function, Disability and Health (ICF). World Health Organization, 2001. For more information on ICF, visit: http://www.who.int/classifications/icf/en/index.html

Note that the ICF framework is not a linear model. Within the ICF model, a child with a health condition such as cerebral palsy may have impairments in body structures (e.g. spasticity). However, this does not necessarily mean that the child will have a participation restriction. While the child may not be able to play on the school basketball team, he can participate in the sport of wheelchair basketball through the local Special Olympics organization. Similarly, while he may not be able to walk, his use of a manual wheelchair affords him mobility and access to his home and community. Nutritionists and other health care professionals have an important role in preventing or minimizing the degree of restrictions in activities and participation, thus minimizing the degree of disability.

Benefits of Physical Activity for Children with Special Health Care Needs

The benefits of physical activity for children with special health care needs have been demonstrated in a growing body of scientific literature, as documented in the studies cited below.

Incorporating regular physical activity into their lifestyles will hopefully increase the likelihood of improved health in adulthood for CSHCN, as well as reduce secondary conditions such as cardiovascular disease and osteoporosis. Equally important are the psychological benefits of physical activities including opportunities to form friendships, express creativity, and develop self-esteem and confidence (3). However, there are important health-related conditions, such as high blood sugar in children with diabetes or cardiac defects in children with Down syndrome, that may be affected by participation in physical activity. Thus, screening and assessment of CSHCN, conducted by appropriate members of the individual's health care team, is recommended prior to participation in physical activity.

Benefits of Physical Activity for Children with Obesity

Obesity is associated with severe health risks (9). In 2015-16, the prevalence of obesity of children and adolescents in the U.S. was 18.5% (10) as defined by a body mass index (BMI) at or above the 95th percentile. Overweight is the term used when one's BMI is between the 85th- 94th percentiles (11). Children with disabilities and special health care needs are more likely to be overweight or obese than their typically developing peers.

There are many societal factors presumed to contribute to the increased prevalence of childhood obesity, including less time playing outdoors and in physical activities, increased "screen" time (tablet, smart phones, video games, TV), increased consumption of high calorie fast foods, and the decline of family meals (12). In addition to an increased risk of obesity in adulthood, there are a number of diseases associated with childhood obesity that can impact one's health for a lifetime. Into adulthood, obese children and adolescents can have associated conditions including asthma,

Type 2 diabetes, hypertension, musculoskeletal complications, psychosocial stigmas, and sleep apnea (13).

In addition to healthy eating habits and decreasing sedentary activities, regular physical activity is a primary strategy for preventing obesity. Of the limited studies on the use of both exercise and nutrition education as an approach to addressing childhood obesity, efficacy of this combination has been reported. An important finding was that an educational approach focusing on exercise and nutrition empowers both children and their families to make better lifestyle choices around activity participation and food selections. Improved lifestyle choices equate with improved health over a lifetime (14).

Benefits of Physical Activity for Children with Asthma

Another group likely to experience limitations in exercise and physical activity is children with asthma, one of the most prevalent disorders in childhood. To avoid the common unpleasant feelings of shortness of breath and exercise-induced bronchospasm (EIB), some children with asthma self-limit their physical activities. Restricting physical activities can contribute to muscle deconditioning and lowered levels of fitness, as well as limit participation in play and recreation with peers.

By preventing or managing EIB, many children and adolescents with asthma can safely participate in physical activities and sports. Prevention and management strategies may include identification and control of triggers (e.g. allergens, prolonged physical activity {especially in cold weather}, respiratory tract infections), medications, and modification of physical activities. Modifications may include longer warm-up and cool-down periods, modifying activity intensity, training and conditioning to prepare for physical activity, and monitoring the environment to minimize asthma triggers. A team approach to asthma management is recommended, with all the adults involved in the care of the child participating. An asthma-management plan should also include the PE teacher at school, as modifications may be necessary for participation and enjoyment.

Research shows that physical activity for children with asthma not only improves fitness, but may also have a positive effect on perceived physical competence and coping with asthma (15,16). Study participants tolerated the training programs well, and led the authors to recommend organized sports activities with short and intense bouts of muscle work for children with stable asthma. Parents of children in the experimental groups reported their children had more success and pleasure in sports, and that they played with other children for longer periods of time (16).

Benefits of Physical Activity for Children with Neuromotor Disabilities

The benefits of physical activity for children with neuromotor disabilities such as Down syndrome and cerebral palsy (CP) have been documented in the literature, as cited below. However, some

children may require a health screening from a primary care provider prior to participating, and some activities may have to be adapted to ensure that the child or adolescent has a safe, positive experience.

Common physical characteristics of children with Down syndrome include hypotonia, decreased muscle strength, and ligamentous laxity, which can limit endurance and restrict participation in physical activities, including play. In addition to these musculoskeletal impairments, there are other associated medical and health issues that can affect physical activity including congenital heart defects, atlantoaxial instability (i.e. an abnormally large space and excessive motion between the first and second cervical vertebrae), and a tendency toward obesity. All health care providers should discuss with families the risks that certain physical activities (i.e. gymnastics, horseback riding) involving neck hyperflexion or hyperextension may pose for individuals with atlantoaxial instability. Parents in turn need to consult with their child's primary care provider before these types of activities are initiated. As long as medical and health issues are monitored, participation in moderate- to vigorous intense activity is recommended for children with Down syndrome to reduce their tendency toward obesity, enhance social opportunities, and promote lifelong health (17).

For many children with CP, impairments such as muscle weakness, muscle spasticity, and balance deficits make it difficult to participate in sport and play activities at sufficient levels to develop and maintain normal physical fitness levels (18). However, a substantial body of evidence has documented that muscle strength can be improved in children with CP, and that improved strength can translate into functional gains such as improved walking efficiency (19, 20). Thus, the child's participation in physical activities should be encouraged by all members of the individual's health care team.

Given the appropriate guidance and supervision, children and adolescents with neuromotor disabilities can improve fitness and physical activity levels at community fitness facilities. This option not only allows these children to participate in community-based settings, but also promotes their involvement in fitness activities with other family members. Programs in the community also offer the possibility of transitioning children from individual physical therapy or occupational therapy sessions to lifelong fitness programs (18). In one study evaluating a conditioning program of aerobic exercise, flexibility exercises and weight training for adolescents with CP, significant improvements in strength were documented (21). However, an unanticipated outcome of the program was a dramatic change in self-perception for the participants, with some participants gaining enough confidence to enroll in a regular aerobics class after the study. This self-initiation represents an important step in health promotion for these adolescents.

Other benefits of community-based activity programs for children with special health care needs are the positive effects on motivation and compliance. Many individuals, including both children and adults, are more motivated to exercise in a group setting. Parents reported difficulties in

getting their young children with physical disabilities to comply with home exercise programs (HEP) (22). But when involved in a group fitness program incorporating strength and endurance training, attendance and adherence to the exercise program was high, prompting parents to request continuation of the program. Improvements in many of the outcome measures such as energy expenditure, strength, fitness, and self-perception were also documented for the study subjects.

The Role of the Nutritionist in Promoting Physical Activity for Children and Youth with Special Health Care Needs

The role of the nutritionist is to collaborate in the development of an overall physical activity plan for the child through the following activities:

- perform screening as indicated in Table 3-1
- provide information regarding the benefits of physical activity and contraindications
- make referrals to other health care professionals when appropriate
- provide appropriate resources
- facilitate partnerships among other health care professionals, families, and community agencies (e.g. schools, Special Olympics)
- serve as an advocate
- provide support and encouragement

One of the most important benefits a nutritionist can provide to these children and their families is anticipatory guidance to help prevent complications of inadequate nutrition and inactivity. Once family activity patterns are identified, guidance about limiting sedentary behaviors (e.g. playing on tablet, watching TV) may be indicated. Education regarding proper nutrition, weight management, and exercise is vital to making choices about a healthy lifestyle. Establishing healthy eating habits and exercise as a part of one's lifestyle when a person is young will help ensure they carry over the behaviors into adulthood.

The following case study illustrates how the nutritionist, using a family-centered approach, helps facilitate a physical activity program for a boy with cerebral palsy.

Charlie is a 10-year-old boy with a history of premature birth at 28 weeks gestation. He has a diagnosis of spastic diplegic cerebral palsy (CP). Charlie is a friendly, social boy, and his cognitive skills are within the average range. He presently ambulates with Lofstrand crutches for short distances at home and at school, but uses a manual wheelchair for community access. The family has recently moved to the area, and Charlie was just enrolled in the 5th grade at a new school. At the first well-child appointment with his new primary care provider, Charlie's growth parameters

met the criteria for obesity. His primary care provider made a referral to the nutritionist at the public health department.

During an interview with Charlie's mother, the nutritionist learned that Charlie's inactivity, coupled with excessive caloric intake, was contributing to his obesity. In addition to his short-distance crutch walking, his primary form of physical activity was limited to a home exercise program (HEP) of stretching developed by his previous physical therapist. Charlie's mother reported that he was not motivated to perform his HEP, and that she was tired of nagging him. She was interested in learning about alternative physical activities that might be more appealing and motivating for Charlie. A follow-up visit was scheduled to design a plan.

At that visit, the nutritionist, Charlie, and his mother collaborated to make a plan to decrease Charlie's caloric intake, as well as increase his physical activity level. The nutritionist discussed some of the questions in Table 3-1 with Charlie and his mother to identify family recreation interests. Charlie was fortunate to be part of a family that valued fitness. The nutritionist learned that Charlie was more motivated to participate in recreation activities with his family than play in adapted team sports. Because of the family's interest in skiing and cycling, the nutritionist suggested contacting Outdoors for All, a local organization providing year-round instruction in outdoor recreation and modified sporting equipment for people with physical, developmental, and sensory disabilities. To help maintain weight and promote overall fitness and conditioning, the nutritionist recommended checking out some of the local fitness clubs for a family membership. She also suggested that Charlie's new physical therapist may be able to consult with the trainer at the club to develop a strengthening and conditioning program for him that would meet the Surgeon General's recommendation for physical activity a minimum of 60 minutes, most days of the week (23).

Screening Guidelines

Children and adolescents with special health care needs vary in their ability to participate in physical activities. Individual screening or assessment should be conducted by appropriate health care professionals, including the nutritionist, before a child begins a program of physical activity. To help the family select appropriate and beneficial physical activities, health care providers need to consider the individual's health status, interests, cognitive skills, and available community resources. Most importantly, working with the family to identify their specific physical activity goals for their child and their current physical activities as a family will help tailor recommendations.

Tables 3-1 and 3-2 provide guidelines for screening of children with special health care needs and appropriate interventions to promote physical activity.

Table 3-1: Suggested Screening Questions About Physical Activity

1. What physical activities does your family currently participate in?
2. What are your child's interests related to physical activity?
3. Does your child participate in physical activities at school? If so, which ones? How often?
4. What are your priorities for your child's participation in a physical activity? (e.g. an activity other family members enjoy, for socialization, weight management)
5. How does your child's health impairment limit his/her participation in physical activities? What type of activities should be avoided?
6. How does your child understand and follow instructions and rules?
7. Has your child had any experience participating in structured group physical activities, such as a gymnastics class, T-ball team, or group setting?
8. What modifications might be necessary for your child to participate in a physical activity? (e.g. adapted equipment, modification of rules, simplified instruction, protective equipment)

Table 3-2: Interventions to Promote Physical Activity in Children and Youth with Special Health Care Needs

Screening/Assessment	Intervention	Evaluation/Outcome
Obtain current medical/health information	Review current medical/health information	Child will participate in activities that are compatible with current health status
Assess nutritional status	Develop a nutrition plan to include physical activity to promote growth and well-being	Child will maximize their nutritional status and participate in appropriate physical activity
Review current medications	Discuss with parents any side effects of medications (e.g. diarrhea, constipation, sleepiness) that may effect physical activity participation	Child will participate in activities that are compatible with medication side effects
Probe for current physical activity levels at home, school (i.e. sports, PE class) and community	Refer to Table 3-1 Educate the family regarding community resources for physical/ recreation opportunities for child Provide guidance re: limiting sedentary activities	Child will participate in physical/ recreational activities geared towards his/her interests and abilities Family will identify and participate in community physical activities with child
Probe for contraindications to physical activity	Discuss with parents and/or contact primary care provider to inquire re: contraindications to physical activities PCP or family member write a cautionary statement to provide to community organizations re: child's limitations and/or contraindications	School and community physical activity providers will have knowledge of child's restrictions and/or adaptations regarding safe and appropriate physical participation Eliminate/minimize possibility of injury for child during physical activity
Inquire if parents have any concerns around child's motor skills and if child is receiving any occupational therapy (OT) or physical therapy (PT) services	Refer to pediatric OT or PT to evaluate motor status and determine if direct OT or PT services are needed to improve motor skills for participation in physical activities	Child will develop necessary motor skills to participate in selected physical activities

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Resources

Ability Path
An online hub and special needs community for parents and professionals to learn, connect and live a more balanced life - through all phases of a child's growth and development.
https://abilitypath.org

American College of Sports Medicine
www.acsm.org/ (go to link: Physical Activity and Public Health Guidelines)

Centers for Disease Control and Prevention
National Center for Chronic Disease Prevention and Health Promotion
www.cdc.gov/nccdphp/dnpa

City/Town Parks & Recreation & Special Populations Programs – Help family locate contact for their local community's park and recreation programs.

Finding Balance: Obesity and Children with Special Needs. Includes many tips and tools for parents.
https://abilitypath.org/wp-content/uploads/2015/11/obesity-report.pdf

Healthy People 2020
https://www.healthypeople.gov/2020/About-Healthy-People

Including all Children: Health for Kids with Disabilities
A national non-profit of volunteers and partners who make schools and communities healthier places for children to thrive. Blog with information specifically for children with disabilities.
https://www.actionforhealthykids.org/blog/page/2/?topicfilter%5B0%5D=Physical%20Activity%2C%20PE%2C%20%26%20Play

The Kid's Activity Pyramid, MU Extension, University of Missouri-Columbia
https://www.mdpedi.com/wp-content/uploads/2015/04/eKids1.pdf

The Kid's Activity Pyramid, Penn State College of Agricultural Sciences Publications,
http://pubs.cas.psu.edu/freepubs/pdfs/uk076.pdf

The National Center on Physical Activity and Disability
https://www.nchpad.org/1747/6895/Active~People~Healthy~Nation

National Physical Activity Plan
http://www.physicalactivityplan.org/index.html

Office of Disease Prevention and Health Promotion
https://health.gov/about-us/

Outdoors for All
https://outdoorsforall.org

Physical Activity for Children and Youth. A 2018 report on physical activity in U.S. children and youth. Chart with Physical Activity Guidelines (See page7).
http://physicalactivityplan.org/projects/PA/2018/2018%20US%20Report%20Card%20Full%20Version_WEB.PDF?pdf=page-link

President's Council on Physical Fitness and Sports
www.fitness.gov/

Special Olympics www.specialolympics.org
Visit this website to learn about Special Olympics. A link is available to find a local Special Olympics program.

Summer Camps and Programs for Children with Special Needs (WA State) - This link is to programs in WA State. Check with local resources for children with special needs to determine if there are summer programs in your area.
https://www.seattlechildrens.org/globalassets/documents/clinics/autism-center/summer-camp-directory-2018.pdf

Surgeon General of the United States
www.surgeongeneral.gov/

TOPSoccer – This is an example of a program in WA State for your review. Check with soccer organizations in your area to determine if programs are available.
The Outreach Program for Soccer (TOPSoccer) is a program for players 4 years of age and older who have developmental and/or physical disabilities.
https://washingtonyouthsoccer.org/community/top-soccer/

United States Dept. of Health and Human Services – "I Can Do It, You Can Do It!"
Nation-wide initiative supporting physical activity for children with disabilities; includes on-line mentoring program with 50 organizations participating
https://acl.gov/programs/health-wellness/icdi

Chapter 4

BREAST/CHESTFEEDING FOR THE CHILD WITH SPECIAL HEALTH CARE NEEDS

Robin P. Glass, MS, OTR, IBCLC and Lynn S. Wolf, MOT, OTR, IBCLC

The nutritional benefits of breast/chestfeeding and human milk for the typically developing infant are undisputed (1). These benefits are equally important for the infant with special health care needs (2). For these babies, there may be additional benefits from ingesting human milk due to their initial medical problems. The anti-infective properties found in human milk often play a crucial role in a baby's recovery from medical complications and support the baby's health in the early months of infancy.

For the typically developing infant, exclusive breast/chestfeeding is considered to be getting all nutrition at the breast/chest without other supplements. For the infant with special health care needs, a broader definition should be considered. Breast/chestfeeding can be seen on a continuum from full nutrition directly from the breast/chest through receiving human milk either fully or partially from a bottle or tube feeding. Attention to the adequacy of the lactating parent's milk supply is crucial for the baby to obtain optimal benefits. In particular, the first few weeks of life are vital to the parent's ability to provide the baby with enough breast/chest milk for growth over the next six to twelve months. All infants who are receiving human milk still need the recommended supplements, including a single shot of Vitamin K at birth, use of a Vitamin D supplement started within a few days of birth, and a supplement of iron introduced in the first few months of life.

If the infant is not able to feed at the breast/chest within one hour after birth, the lactating parent should be assisted to begin pumping using a hospital grade pump. The first two weeks after birth are a crucial time for the establishment of a full milk supply (3).

Encourage the parent to ideally pump a minimum of eight times per day, spaced at 2-3 hour intervals during the day with a break of no longer than 4-5 hours at night. The aim is to produce at least 16 oz or more of breast/chest milk per day at two weeks after birth.

Pumped expressed breast/chest milk can be stored for four days in the back of the refrigerator or frozen and kept for 3-6 months in the freezer of a refrigerator with a separate door for the freezer. If the lactating parent is struggling with their milk supply even though they are pumping regularly,

a referral to a lactation consultant (LC) is indicated. The LC, in conjunction with the parent's primary medical doctor, (PMD) might recommend galactogogues to help boost milk supply. These are medications and herbs that can increase the lactating parent's rate of milk production and help their pumping efforts (1). The LC may also evaluate the type of breast/chest pump the parent is using, the fit of the breast/chest pump flanges, and the amount of suction the parent is using to identify optimal pumping technique.

Many lactating parents worry that their baby is not getting enough milk at the breast/chest. This can sometimes reflect a new parent's lack of confidence, but it can also be an indication that their milk supply is low or that the baby is having trouble transferring milk from the breast/chest (4). If the baby has inadequate growth, falls asleep quickly after starting to nurse, or appears to be hungry even after feeding, a detailed assessment is indicated to identify the problem. Issues with breast/chestfeeding can reside with the lactating parent, infant or both. Careful evaluation by a registered dietitian nutritionist, a lactation consultant, and an occupational, physical or speech therapist with a specialty in breast/chestfeeding can determine the problem and develop treatment strategies.

The most common reasons lactating parents have low milk supply are related to incomplete milk removal by the baby, delay in the start of pumping after birth if the baby is unable to feed at the breast/chest, and/or low frequency of pumping (3). Lactating parents may also have hormonal issues such as hypothyroidism, retained placental parts, or hypoplastic breast/chest development during pregnancy that can be linked with poor milk supply (3).

Once the contributions from the lactating parent-sided issues are determined, a more detailed evaluation of the infant's role can occur. Assessment of the infant's oral motor control as it relates to the ability to latch to and remove milk from the breast/chest is an important first step (5). When observing a lactating parent or baby breast/chestfeeding, a pre-post breast/chestfeeding weight using an appropriate gram scale is the only reliable method to determine intake from the breast. The amount of time spent at the breast/chest is an extremely inaccurate measure of milk transfer. In addition to pre/post weights, observing the infant's sucking pattern and rate of swallowing can give some indication of the baby's efficiency of milk transfer. Frequent weight checks to monitor overall weight gain and growth velocity will also provide valuable data on which to base a treatment plan.

How satisfied the baby appears after nursing and the length of time between feedings can also provide clues to the adequacy of milk transfer from the lactating parent to the infant. However, behavioral cues alone may not accurately reflect the amount of nutrition the baby is receiving at the breast/chest. Infants with special health care needs may be particularly vulnerable to under-eating, as they may have diminished endurance from their primary medical conditions.

Many infants will require additional calories beyond what they are capable of taking each day

in order to grow adequately. The RDN's individualization of the care plan for each infant and lactating parent should be done in a manner that ultimately leads to full breast/chestfeeding and/or use of the parent's milk supported by methods to preserve the milk supply. The energy density of a lactating parent's expressed breast/chest milk (EBM) can be increased by the use of powdered formula and/or additional additives. See Appendix F. The increased energy dense breast/chest milk can be given by a supplemental nursing system (SNS) or by use of the bottle for 1 or 2 feedings a day with the goal of full breast/chestfeeding and/or use of all breast/chest milk. Careful evaluation of the infant's growth will be needed. Merely taking the fully breast/chestfed baby off of the breast/chest, having the lactating parent pump milk, fortifying it, and then giving it by bottle can quickly lead to the cessation of breast/chestfeeding, and possibly a severe reduction in breast/chest milk supply.

Babies who require nasogastric or gastrostomy tube feedings can also obtain breast/chestfeeding benefits. They may breast/chestfeed for a portion of their nutrition with tube feeding volumes adjusted to account for intake (as measured by pre-post weights).

The goal would be for babies to take as much EBM as possible through tube feedings. Babies who take low volumes from the breast/chest or who are unsafe to breast/chestfeed can still nurse at a pre-pumped ("dry") breast/chest. These babies should continue to participate in skin-to-skin care as these experiences can have a beneficial influence on milk production and bonding between parent and baby.

Contraindications for breast/chestfeeding and/or use of human milk are occasionally present in babies with special health care needs. The most obvious is for infants identified with galactosemia or other inborn errors of metabolism (See Chapter 19). For other contraindications to breast/chestfeeding and/or the use of human milk see The American Academy of Pediatrics Pediatric Nutrition Handbook (6).

For the infant with special health care needs, breast/chestfeeding may look different for each parent/baby pair. The primary goal is for the baby to receive as much human milk as possible, with the secondary goal of achieving some feeding at the breast/chest. Treatment strategies must support the lactating parent in maintaining their milk supply and should support the parent and baby in moving toward breast/chestfeeding. The intensity of the physical and emotional experience for the lactating parent beginning breast/chestfeeding with an infant with special health care needs should be acknowledged and support provided. The team of professionals working together can help balance competing medical goals, provide emotional support, and offer practical guidance to ensure the baby's optimal growth. In the process, we may redefine "breast/chestfeeding" in a way that is unique to each parent/baby pair.

Table 4-1 presents guidelines for the assessment, intervention, and outcome/evaluation for several breast/chestfeeding concerns.

Table 4-1: Nutrition Interventions for Breast/chestfeeding

Assessment	Intervention	Evaluation/Outcome
Is breast/chest milk supply adequate?		
A lactating parent may have low milk supply if any of the following are observed: • Infant has inadequate weight gain or slow growth velocity • Insufficient number of feedings or length of feeds/day • Pumping volumes are low • Inadequate milk transfer based on pre/post breast/chestfeeding weights • History of no change in breast/chest size prenatally or within 1-2 weeks after birth • Infant appears hungry after feeding or eats more often than every 2 hours • Infant requires supplemental formula feeds for growth	If low milk supply is observed, refer lactating parent to a lactation consultant (LC) for further evaluation and intervention. Lactating parent should begin pumping with a hospital grade pump, at least 8-10 times per day. Consider beginning a galactagogue	Infant will demonstrate age appropriate growth. Lactating parent will maintain milk supply
Is milk transfer effective?		
In a lactating parent with good milk supply, if latch is effective there should be clear evidence of appropriate milk transfer, such as: • Active sucking throughout most of a 10 -20 minute period at one or both breasts/chest (baby should not be mostly sleepy) • Most sucking with "long draws" (not short, rapid sucks) • Evidence of swallowing after every 1-2 sucks • Baby satisfied when comes off of the breast/chest • Pre-post weights using digital scale (accurate to +/- 2 grams) show adequate intake. • There should not be significant pain Remember: the amount of time spent at the breast/chest is not an accurate indication of the amount of milk the baby is getting	If milk supply is clearly good (high pumping volumes), but baby is not transferring milk effectively: **Improve latch:** • Optimize position • Asymmetric latch techniques **Referral to LC if:** • Changes to position/latch not helping • Suspect problem with baby's sucking • Lactating parent has significant pain	Baby receives adequate nutrition from breast/chestfeeding for optimal growth

Should breast/chestfeeding be supplemented?		
If the baby is not gaining adequately on exclusive breast/chestfeeding, there is other evidence of inadequate milk transfer, or the baby's medical condition requires it, supplementary feeding should be provided. The amount and type will be based on: • Lactating parent's current milk supply • Baby's current success with breast/chestfeeding and milk transfer • Prediction of future potential for breast/chestfeeding • Baby's medical condition The primary goal should be as much nutrition as possible from breast/chest milk, with the secondary goal of successful feeding at the breast/chest	Schemes for providing supplementation: **Baby is primarily non-orally fed:** • Tube feeding required • Maximize amount of breast/chest milk provided • Support skin to skin care (kangaroo care) • Support non-nutritive or low volume nutritive experience at the breast/chest; support from LC as needed **Baby is partially orally fed:** • Tube feeding as necessary. • Breast/chestfeeding based on baby's ability (for nutrition versus for practice); support from LC as needed. • Use of other oral supplementing devices as needed (see below) • Maximize use of expressed breast/chest milk. **Baby is fully orally fed:** • As much nutrition as possible at the breast/chest (balance needs of baby, lactating parent, and family) • Work with LC to build breast/chestfeeding skills • Use of appropriate oral supplementing devices (see below) to support current goals and needs. • Maximize use of expressed breast/chest milk given by supplementary method **Oral supplementing devices can include:** • Finger feeding • Tube and syringe or commercial supplementer at breast/chest (SNS, Hazel Baker finger feeder or Lact- Aid) • Binky trainer • Bottle (select one to provide characteristics similar to breast/chestfeeding, if possible)	Baby receives adequate nutrition using a variety of methods that provide as much breast/chest milk as possible, and work toward breast/chestfeeding as is desired by the family and seems appropriate for the baby

continued...

Assessment (continued)	Intervention	Evaluation/Outcome
Is fortification needed?		
Babies may require fortification to provide extra calories for growth if: • The baby is not able to take adequate volume at the breast/chest, or with human milk + supplement • There are additional caloric or nutritional requirements specific to the baby's medical condition.	If fortification is needed, try to rely on breast/chestfeeding and/or human milk as much as possible: • During pumping, separate hind milk, and use this as a supplement. • Add energy enhancement to expressed breast/chest milk • Balance feedings at the breast/chest with energy enriched bottle feedings • Breast/chestfeed using energy enriched human milk through a tube feeding device (i.e. supplemental nursing system)	Infant will show growth in all parameters at an appropriate rate

Resources

1. Wambach, K, Spenser, B. *Breastfeeding and Human Lactation*, 6th ed. Sudbury, MA: Jones & Bartlett. 2021.

2. Lawrence RA, Lawrence R M. Breastfeeding: *A Guide for the Medical Professional*, 9th ed. Maryland Heights, MO: Elsevier Science. 2021.

3. Marasco, L. West D, *Making more milk*, 2nd ed. New York, NY: McGraw-Hill. 2020.

4. Mohrbacker N. *Breastfeeding Answers: a guide for helping families*. 2nd ed. Preclarus Press, 2020.

5. Genna C W. (editor). *Supporting Sucking Skills in Breastfeeding Infants*. 3rd ed. Sudbury, MA: Jones & Bartlett. 2017.

6. Kleinman RE, Greer FR (eds). *Pediatric Nutrition*. 8th ed. Itasca, IL: American Academy of Pediatrics. 2019.

Chapter 5

MEDICATION-NUTRIENT INTERACTIONS

Starla Blank, RPh, PharmD and Eileen Harper, Med, RD, CD
Updated by Lori Brizee, MS, RD, LD, CSP

Medications and nutrients are known to interact, sometimes with detrimental effects. Medications can affect nutritional status in the following ways:

- Altering the absorption, metabolism, and/or excretion of specific nutrients
- Causing gastrointestinal disturbances and/or anorexia, thereby decreasing overall nutrient intake
- Increasing appetite, which can contribute to overnutrition
- Interacting with nutrients prior to ingestion, such as when mixed with food or formula before administering

Additionally, specific foods and nutrients are known to interfere with the action of certain medications by altering the absorption or metabolism of the medication. Children and youth with special health care needs are at risk for medication-nutrient interactions, especially when medications are used long-term, multiple medications are prescribed, and nutrient intake is marginal (1). Other issues to consider include interactions between medications and vitamin and mineral supplements and the timing of medication administration relative to meals and snacks. This section discusses nine types of medications that have documented effects on nutrients and are commonly used in the treatment of children and youth with special health care needs (1, 2, 3, 4).

Anticonvulsants

Children and youth with neurologic impairments often have secondary seizure disorders that are treated with anticonvulsant medications. Long-term use of anticonvulsant medications places a child at risk for deficiencies of vitamin D, folic acid, and possibly other micronutrients, including vitamin B12 and carnitine (5-13). Routine assessment for vitamin deficiencies is an important component of comprehensive health care for these patients.

Vitamin D

Long-term use of anticonvulsants has been associated with vitamin D deficiency, resulting in rickets or osteomalacia (5-11). The effects of anticonvulsant therapy on vitamin D status are

multiplied by the following factors (5-11):

- Use of anticonvulsant medication therapy for over two years
- Use of enzyme-inducing medications
- Female sex
- High BMI
- Use of multiple medications
- Inactivity or non-ambulatory status
- Little exposure to sunlight
- Dark skin pigmentation
- Inadequate nutrition in general, especially low intake of vitamin D

The anticonvulsants most frequently implicated in vitamin D deficiency are phenytoin (Dilantin), phenobarbital, and carbamazepine (Tegretol). Primidone (Mysoline), valproic acid (Depakene/Depakote), lamotrigine (Lamictal), clonazepam (Rivotril, Klonopin), gabapentin (Neurontin), topiramate (Topamax), and ethosuximide (Zarontin) have also been shown to be associated with vitamin D deficiency and decreased bone mineral density (1,5-11).

Vitamin D levels must be evaluated, and any deficiencies should be treated at the time of initiation of anticonvulsant therapy and regularly thereafter. Children and youth on anticonvulsant therapy who have normal vitamin D levels should be given a prophylactic dose of up to 2000 IU vitamin D daily. Those with documented deficiencies will need pharmacologic doses prescribed by their physicians (1).

Folic Acid, Vitamin B12 and Homocysteine

Low folic acid and vitamin B12 levels with associated high homocysteine levels are seen with most first generation anticonvulsants (e.g., carbamazepine (Tegretol) and phenytoin (Dilantin)). Recent research suggests that folate metabolism is also affected by some of the newer anticonvulsants (e.g., lamotrigine (Lamictal), levetiracetam (Keppra), and zonisamide (Zonegran). Folic acid deficiency is associated with hyperhomocysteinemia, which in turn increases the risk of heart disease. Supplementation with folic acid has been shown to both replete folic acid levels and decrease homocysteine levels in individuals on anticonvulsants (12, 13).

Carnitine

Numerous studies have shown that plasma carnitine levels are significantly lower among patients taking valproic acid than among controls (14). Carnitine deficiency in epilepsy results from a variety of etiologic factors including underlying metabolic disease, inadequate nutrient intake, and specific medication effects. The relationship between carnitine deficiency and valproic acid-induced hepatotoxicity is unclear. Carnitine treatment does not always prevent the emergence of serious hepatotoxicity, but it does alleviate valproic acid-induced hyperammonemia (14, 15).

Medications for Treatment of Attention-Deficit/ Hyperactivity Disorder

Attention-deficit/hyperactivity disorder (ADHD) is commonly treated with stimulant medications such as methylphenidate (Ritalin, Ritalin SR, Metadate CD, Metadate ER, Concerta), dextroamphetamine (Dexedrine), and amphetamine mixture (Adderall). Automexetine (Strattera) is a non-stimulant drug used to treat ADHD. Studies have shown that all of these medications, especially stimulant medications, are associated with decreased appetite in children, often resulting in a slower rate of weight gain and growth. (17, 18) Additionally, a recent cross-sectional study reported an association between ADHD medication usage and decreased bone mineral density among children aged 8-17 years (19). Using NHANES data, Howard et al. showed significant differences in bone mineral density for children and adolescents taking ADHD medications as compared to similar children not taking any prescription medications. (19) In part, this may be influenced by poor dietary intake of total energy, calcium, and vitamin D in children with ADHD who are treated with medication (19). A 2017 study by Bowling et al. showed that children on ADHD medications had decreased BMI between 8 and 11 years of age; however, continued ADHD medication use through 14 years of age correlated with an increased BMI trajectory (20). By 14 years of age, BMI in treated patients was not different from typically developing children or those with ADHD who were not on medications. The authors concluded that a tendency toward overweight based on BMI may result with long term use of ADHD medications after 11 years of age. The authors recommended that lifestyle counseling be initiated following an ADHD diagnosis, regardless of weight status, as children with ADHD are at high risk of poor nutritional intake, irrespective of medication use (20).

Diuretics

Diuretics are frequently prescribed for children with cardiac defects or chronic lung disease. Many diuretics, such as furosemide (Lasix), increase the excretion of potassium, calcium, sodium, zinc, chloride, and magnesium. Potassium-sparing diuretics, such as spironolactone (Aldactone), increase the excretion of calcium and magnesium (3). The diets of patients on diuretics must provide adequate replacement of the minerals that are excreted. Diuretics can also contribute to anorexia and gastrointestinal distress (3,16).

Corticosteroids

Glucocorticoids are used as replacement therapy in adrenocortical deficiency states and for anti-inflammatory and immunosuppressive effects in the treatment of many disorders, including asthma. Side effects of glucocorticoids include decreased bone mineral density; decreased absorption of calcium and phosphorus; poor linear growth; increased appetite often leading to excessive weight gain; sodium and water retention occasionally leading to hypertension; muscle catabolism; increased plasma glucose concentration leading to insulin resistance; and

increased lipolysis (21-25). Inhaled corticosteroids are now more commonly used than systemic corticosteroids. Their side effects are minimal compared to oral corticosteroids, but they can have similar effects when used long term.

Psychotropic Medications (other than ADHD medications)

Psychotropic drugs include antipsychotics, antidepressants, anti-anxiety medications, mood stabilizers, and stimulants used to treat various psychiatric disorders. Several psychotropic drugs used to treat psychotic disorders tend to cause weight gain, especially second generation anti-psychotics (e.g., risperidone (Risperdal), olanzapine (Zyprexa), and quietiapine (Seroquel)). These medications also appear to increase the risk of hyperlipidemia and type 2 diabetes.(26) Metabolic monitoring of weight, lipid profile, blood glucose, and blood pressure is recommended beginning at the start of treatment. A 2017 study by Kauffman et al. showed that monitoring at baseline and at follow-up is not routinely done in young children prescribed second generation anti- psychotic medications.(27)

See Table 5-1 for assessing medication-nutrient interactions for antidepressants (e.g., tricyclic antidepressants (TCAs) and selective serotonin reuptake inhibitors (SSRIs)) and anti-anxiety medications.

Weight management in children and youth with mental health disorders who are taking psychotropic medications needs to be tailored to the individual; for example, the approach to weight management for an individual with obsessive compulsive disorder (OCD) will be different than the approach for an individual with depression. A child with OCD needs a very structured approach to weight management; they may need guidelines regarding specific amounts of different foods to eat to avoid overeating a food labeled as “good” or “healthy” or totally avoiding something perceived as “unhealthy.” The child with depression may benefit from motivational interviewing to come up with ways to increase physical activity or decrease caloric intake. The RDN needs to work closely with the child’s family and mental health professionals to determine the best approach. Optimally, the risk of excessive weight gain needs to be addressed at the time a medication is started so that preventative measures can be taken.

Antibiotics

Antibiotics are used to treat bacterial infections. They are sometimes used long-term on a prophylactic basis. Side effects that may interfere with an adequate nutrient intake include mouth and tongue sores, diarrhea, nausea, and vomiting (16). With long-term use in children with severe disabilities, gut flora can be altered, and vitamin K production may be decreased, increasing the risk of bleeding (28). Probiotics may be used for the prevention and treatment of antibiotic-

associated diarrhea, though current research does not provide specific dosage recommendations (29). Monitoring of nutritional effects is indicated.

Anti-Inflammatory Medications

Anti-inflammatory medications (e.g., sulfasalazine (Azulfidine) for ulcerative colitis and Crohn's disease) can cause nutrition-related side effects including anorexia, nausea, vomiting, taste changes, diarrhea, gastric distress, and abdominal discomfort (3, 16).

Anti-Gastroesophageal Reflux Disease Medications

These medications are used to treat heartburn due to gastroesophageal reflux disease (GERD) by several mechanisms, including increasing GI motility and suppressing acid release in the stomach. Acceleratedgastric emptying may affect the rate of absorption of other medications and lead to a wide range of nutritional side effects, including constipation, diarrhea, nausea, vomiting, and abdominal pain and discomfort (1,2,3). Acid-suppressing proton-pump inhibitors (e.g., omeprazole (Prilosec) and lansoprazole (Nexium)) and H2 blockers (e.g., famotidine (Pepsid) and cimetidine (Tagamet)) may decrease the absorption of micronutrients, such as vitamin B12 and iron, which require a low pH for absorption (3).

Antispasmodics

Antispasmodic medications suppress gastric muscle spasms and may be used to treat conditions like bladder instability, e.g., with myelomeningocele; tone, e.g., with spasticity; and inflammatory bowel syndrome. Some of the nutrition-related adverse effects include nausea, dry mouth, constipation, abdominal pain, anorexia, dysgeusia, and difficulty swallowing (3, 16).

The remainder of this chapter presents guidelines for nutrition assessment, intervention, and evaluation/outcome for specific medication-nutrient interactions.

Table 5-1: Medication-Nutrient Interactions

Assessment	Intervention	Evaluation/Outcome
In addition to the Nutrition Assessment described in Chapter 1, complete the assessment indicated below	See the intervention below for each type of medication	
ANTICONVULSANTS Examples: Any of the following alone or in combination with other anticonvulsants: phenobarbital; phenytoin (Dilantin); carbamazepine (Tegretol); primidone (Mysoline); valproic acid (Depakene/Depakate)		
Assess diet for overall nutrient intake. Check specifically vitamin D, folic acid, other B vitamins, vitamin C, vitamin K, and calcium Monitor for weight gain, weight loss, diarrhea, and constipation	If intake of any nutrient is less than the DRI/RDA: • Counsel caregiver/patient about food sources of nutrients • Recommend multivitamin/mineral supplement at DRI/RDA levels • Monitor for weight changes	Dietary intake of all nutrients is adequate
Assess indicators of bone mineralization: • Serum 25 (OH) vitamin D • Serum calcium (Ca) • Serum phosphorus (P) • Serum alkaline phosphatase (Alk Phos)	If vitamin D is low, supplement with pharmacologic doses of vitamin D3 until it is well within normal limits While pharmacologic doses of vitamin D are being given, monitor for vitamin D toxicity weekly or bimonthly by checking serum Ca level (High serum Ca may indicate vitamin D toxicity) If vitamin D is normal, give up to 2000 IU/day of vitamin D to maintain stores If Ca or P is low, and vitamin D is normal, supplement with Ca or P. If Ca or P is low and vitamin D is low, ensure DRI levels of Ca and P while vitamin D is being repleted If serum Ca, P, and Alk Phos are normal, re-assess 1-2 times per year	Indicators of bone mineralization are within normal limits
STIMULANTS Examples: methylphenidate (Ritalin); dextroamphetamine (Dexedrine, Adderall); pemoline (Cylert)		
Assess dietary intake when medication first prescribed	If diet is low in any nutrient, counsel appropriately. Instruct caregiver(s) to offer meals before giving the medication and later in the day when the medication action is minimal or absent. Collaborate with school to make sure child gets meal or snack before medication at school, if appropriate (see Chapter 12)	Dietary intake of all nutrients is adequate
Assess growth (height or length and weight) every 3 months	If rate of growth (height/length, or weight) is slowing, re-assess dietary intake and counsel appropriately	Weight and height (or length) are increasing in appropriate percentiles

Reassess dietary intake with a 3-to-7-day food record (if possible) and a diet history	If diet is adequate, but growth rate continues to slow, refer to physician to evaluate need for a change in medication or dose	
DIURETICS Examples: furosemide (Lasix); spironolactone (Aldatone); triamterene (Dyrenium)[1] ; thiazides (Diuril, Hydrodiuril, Naqua, Hygroton, Hydromox, and Diamox)		
Consider effect of diuretic on excretion of potassium (K), magnesium (Mg), and calcium (Ca) Assess diet for K, Ca, and Mg	If intake of K, Ca, or Mg is lower than the DRI, counsel regarding dietary sources Consider mineral supplements of Mg and Ca to meet the DRI.	Dietary intake of all nutrients is adequate
If use of diuretics has been long-term, assess electrolyte and mineral status	If mineral deficiency is evident, counsel on dietary sources and provide mineral supplement	Lab indicators of electrolyte and mineral status are within normal limits
CORTICOSTEROIDS Examples: **Systemic** - dexamethasone, hydrocortisone, methylprednisolone, prednisolone, prednisone **Inhaled** - triamcinolone acetonide (Azmacort)		
Assess for indigestion or mild GI intolerances that may occur Assess if patient receiving prolonged therapy with pharmacologic doses	Administer oral or inhaled dosage forms with food Consider need for sodium restriction and/or potassium supplementation	GI distress is decreased Fluid retention and electrolyte disturbances are minimized
Assess protein intake to ensure adequacy	Ensure adequate intake of protein and encourage physical activity	Muscle catabolism is minimized.
Assess vitamin and mineral intake	Supplement any vitamins and minerals that are deficient in diet Supplement calcium to insure intake of 150% DRI22 and 400 to 2000 IU vitamin D	Intake of vitamins, calcium, and phosphorus is adequate Assess linear growth
Assess linear growth	Discuss possibility of decreased dose and/or alternate days on/off medication with physician	Effect on growth is minimized
Refer to PCP to assess bone density—use bone densitometry to diagnose osteoporosis if long term corticosteroid treatment	If condition permits, exercise or physical therapy will reduce risk of osteoporosis	Bone loss is prevented/minimized
ANTIDEPRESSANTS Examples: **Tricyclic antipressants (TCAs)** - amitriptyline (Elavil); amoxapine (Asendin); clomipramine (Anafranil); desipramine (Norpramin); imipramine (Tofranil); nortriptyline (Aventyl, Pamelor); protriptyline (Vivactil); trimipramine(Surmontil); **Selective Serotonin Reuptake Inhibitors (SSRIs)** - fluoxetine (Prozac); sertraline (Zoloft)		
Assess if patient on tricyclic antidepressants (TCAs) or selective serotonin receptor inhibitors (SSRIs)	Monitor for dry mouth, taste changes, GI distress Take in morning without regard to meals Monitor weight	Decreased GI distress. Appropriate rate of growth and weight gain
Assess if problem with gastric irritation	Take medication with or immediately after food to lessen irritation (for TCAs)	Decreased GI distress

continued...

Assessment (continued)	Intervention	Evaluation/Outcome
Assess if patient on amitriptyline (Elavil) or imipramine (Tofranil)	Requirements for riboflavin may be increased/ may interfere with the biochemical assessment of riboflavin's effect or induce riboflavin depletion	Adequate riboflavin intake
ANTI-ANXIETY Examples: diazepam (Valium)		
Monitor for dry mouth, nausea, constipation, hypoalbuminemia (with usage over 4 weeks)	Increase fluid intake as needed Check serum albumin every 6 months Ensure protein needs are met; modify protein intake as needed	Serum albumin is within normal limits Problems with dry mouth, constipation are minimized
ANTIBIOTICS Examples: cefazolin (Ancef, Kefzol); cefotaxime (Claforan); cefotetan (Cefotan); ceftizoxime (Cefizox); ceftriaxone (Rocephin); penicillin; vancomycin		
Assess for anorexia or GI distress	Anorexia – Suggest small, frequent meals GI distress - If appropriate, suggest medication be taken with meals Consider probiotics to prevent or treat antibiotic-associated diarrhea	Rate of growth and weight gain is appropriate GI distress is minimized
ANTI-INFLAMMATORY Examples: sulfasalazine (Asulfidine)		
Assess for GI distress Assess fluid intake Assess folate intake	Take with water after meals or with food Ensure adequate urine output (> 1500 cc/day) Folate supplement (1 mg/day) recommended Take folate separately from medication	Decreased GI distress Sufficient urine output Prevent folate deficiency
ANTI-GASTROESPHAGEAL REFLUX DISEASE (GERD) Examples: ranitidine (Zantac) **Proton-pump Inhibitors** - omeprazole (Prilosec), lansoprazole (Prevacid), esomeprazole (Nexium) **H_2 Blockers** - famotidine (Pepsid), cimetidine (Tagamet), nizatidine (Axid)		
Assess for GI distress	Take with meals and/or bedtime snack Bland diet may be recommended Limit caffeine	Decreased GI distress
Assess vitamin and mineral intake, especially iron, calcium, magnesium, and zinc	Recommend dietary sources for any deficient micronutrients Supplement any vitamins and minerals that are deficient in diet	Intake of vitamins and minerals is adequate
ANTISPASMODICS Examples: oxybutynin (Ditropan), tizanidine (Zanaflex), baclofen (Lioresal), dantrolene[2] (Dantrium)		
Assess for GI distress	Take consistently with or without food (food increases maximum concentration and decreases time to peak concentration)	Decreased GI distress
Assess for swallowing difficulty	Refer to PCP/SLP for evaluation Modify diet texture/thickness as needed	Improved oral intake and ability to meet daily needs

[1] Spironolactone and triamterene are potassium-sparing diuretics. Avoid salt substitutes that are high in potassium. Do not supplement with potassium unless serum K low and only under close supervision.
[2] With dantrolene, monitor for GI bleeding and dysphagia

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CHAPTER 6

NUTRITION INTERVENTIONS FOR CONSTIPATION

Katrina Hoch, PhD, MS, RD

Many children and youth with special healthcare needs are particularly susceptible to constipation, defined as infrequent, difficult, painful, or incomplete evacuation of stools. Constipation is also understood to include passage of hard, small, pellet-like stools or large diameter stools (1, 2, 3, 4). Definitions vary, but most clinicians and guidelines agree that a stool frequency of less than three times per week would be considered constipation in any age group (1, 2, 3). However, infrequent stooling is not required for a diagnosis of constipation.

Children experiencing constipation may present with abdominal pain, abdominal distention, decreased oral intake or fecal incontinence. If undiagnosed or under-treated, constipation can lead to significant complications including anal fissures, urinary tract infections, appetite suppression, growth failure, emotional distress, family disruption, and educational difficulties. Constipation can also create a cycle of mutually reinforcing behavioral stool withholding and impaction. As stool sits in the rectum, water is absorbed into the intestinal walls and the stool becomes harder, larger, and more difficult and painful to pass. This can lead to withholding as children seek to avoid stooling and the associated discomfort. Chronic constipation can cause fecal impaction and rectal distention, and can result in overflow soiling, or encopresis, around the impacted stool mass (1, 3, 4). Constipation can also lead to increased health care costs and lower quality of life (2, 4, 5, 6). Inadequate or delayed treatment for pediatric constipation is associated with long-term constipation in adulthood (2, 4).

Constipation can be the result of an organic disease process, or it may be considered functional constipation, defined as constipation that is not explained by an underlying anatomical or physiological condition (1, 2, 3). The etiology of functional constipation is multifactorial, but stool withholding to avoid pain is often a contributor (1).

Several factors predispose children to functional constipation. These include psychological stress, physical or emotional maltreatment, sexual abuse, and lack of consistent daily routine (4, 6). Additionally, certain life transitions such as starting solid foods or cows milk, starting toilet training, changing housing, and starting daycare or school frequently become precipitating triggers (2, 6).

Table 6-1: Constipation Causes and Contributors in Children and Youth with Special Health Care Needs (1-4, 6, 7)

Lifestyle/Psychosocial	Medical/Developmental
Dietary issues (e.g., inadequate fiber intake, diet composed primarily of highly processed foods) Inadequate fluid intakes Low level of physical activity due to sedentary lifestyle Excessive cow milk intake Impaired mobility (e.g., due to prolonged illness or injury, impaired motor skills, paralysis) Unable to communicate need Behavioral withholding Coercive toilet training or struggles around toilet training Lack of routine toileting habits or inability to attain upright position Inadequate intake or malnutrition (e.g., in underfeeding, anorexia nervosa, bulimia nervosa)	Conditions associated with atypical sensory processing (e.g., autism spectrum disorder and attention-deficit/ hyperactivity disorder) Cow milk protein allergy or intolerance Excessive fluid losses (e.g., due to drooling, chronic vomiting, or fever) Medications (e.g., opiates, anticholinergics, antidepressants, chemotherapy) Anatomic anomalies (e.g., imperforate anus, anal stenosis) Abnormal neurologic function of the intestinal tract (e.g., Hirschsprung's disease, anal achalasia, chronic intestinal pseudo-obstruction, neurogenic bowel associated with spinal cord anomalies) Abnormal musculature or muscle tone leading to impaired intestinal function (e.g., prune belly syndrome, gastroschisis, Down syndrome, cerebral palsy, Duchenne muscular dystrophy). Metabolic or endocrine conditions (e.g., hypothyroidism, hypercalcemia, hypokalemia) Cystic fibrosis Celiac disease Multiple endocrine neoplasia type 2B Sacral teratoma/tumor Vitamin D toxicity Heavy metal poisoning Infant botulism

Children with constipation require prompt treatment. Delaying treatment for more than three months beyond onset of symptoms is associated with longer duration and increases the risk of starting a cycle of pain, withholding, and impaction (2, 3, 4).

Treatment course will depend on the age and developmental stage of the child, whether the constipation is recent onset or chronic, and the precipitating triggers or organic cause of constipation. The plan will generally consist of treating the organic disease if present, dietary counseling to increase fiber and fluids, increased physical activity, routine toileting schedule and proper positioning, parent education about toilet training, behavioral intervention, and use of

laxatives (2, 6). If impaction is present, disimpaction is accomplished with higher doses of laxatives, suppositories, or enemas (depending on the child's age). A list of laxatives and description of mechanisms is provided in Table 6-2. The goal of treatment is to produce one to two soft stools per day.

Several areas of treatment for constipation that have sparked some controversy are discussed below.

High Fiber Diet

Children and youth with special healthcare needs suffering from constipation may benefit from increasing their dietary fiber intake. High fiber diets for children have been a subject of debate due to concerns that they reduce energy intake and the bioavailability of vitamins and minerals. Studies have produced mixed results regarding whether this is the case in practice (8, 9). Observational studies show an association between low fiber intake and pediatric constipation, though these studies do not establish causality (10, 11, 8). Evidence also indicates that most children do not meet fiber recommendations with a majority receiving less than 50 percent of the Institute of Medicine's Dietary Reference Intakes (8, 10, 12, 13). Fiber intake may be lower for children and youth with special healthcare needs who have feeding challenges. Thus, increasing fiber intake will often mean bringing it closer to recommended levels rather than exceeding them. The goal in most cases is adequate fiber rather than high fiber (10).

Recommendations for children's fiber intake vary. The 2020-2025 Dietary Guidelines, based on the Dietary Reference Intakes (DRIs), use the goal of 14 g/1000 kcals to provide recommendations based on age and sex (14). The American Academy of Pediatrics (APP) recommends using age in years + 5 g/day or 0.5g/kg body weight up to 35 g/d (8, 10). These goals may not fit the precise needs of children and youth with special healthcare needs who are on low energy diets, tube feeding diets, or other specialized regimens; these children may need additional fiber. The AAP recommends staying below age + 10 g/day (10). There are no fiber recommendations for infants below one year of age (8, 9).

Guidelines from many organizations recommend normal fiber intake through a balanced diet with adequate fluids as part of the overall treatment plan for constipation (2, 3, 9, 13). Increasing fiber through whole foods such as fruits, vegetables, whole grains, legumes, and nuts is ideal, as many whole plant foods contain multiple types of fiber and additional compounds such as sorbitol that contribute to healthy stooling patterns. They also help maximize the nutritional value of the diet (8, 9). Children fed enterally may benefit from having home-blended foods, commercial blended food formulas, or formulas with fiber added to their regimen.

For children who cannot consume fiber-rich whole foods, fiber supplements are sometimes used. However, there is insufficient evidence to support a recommendation for many of the

available fiber supplements (2, 9, 10, 12, 13). Fibers differ in properties and mechanisms for laxation, and not all fiber types improve bowel function (9, 11, 12). To help with stool output, a fiber must at least partially resist fermentation and remain intact throughout the colon (11, 12). Large, coarse particles of insoluble fiber, such as coarse wheat bran, mechanically irritate the colonic mucosa, stimulating secretion of water and mucus and increasing stool water content (11, 12, 13). Soluble, gel-forming fiber, such as psyllium, has a high water-holding capacity and resists stool dehydration. Both of these mechanisms lead to softening and bulking stool, making it easier to pass. Clinical evidence supports the use of coarse wheat bran and psyllium for treating constipation (11, 12). Additional water should be provided when using a fiber supplement, as more water will be lost in the stool (8, 9).

Dietary changes should be used in concert with the overall treatment plan, including laxatives, behavioral modifications, and other therapies as needed. When constipation is the result of organic disease, it is particularly important to discuss the plan with the medical team and to address the organic cause (2, 3). Assess each child's diagnoses and consider whether increased fiber is appropriate. Use caution with obstructive, metabolic, neurological, and myogenic constipation etiologies. For patients with irritable bowel syndrome (IBS), particularly if they are following a low FODMAP diet, increasing insoluble fiber may worsen symptoms (11, 13). When increasing fiber intake, do so gradually over a period of weeks and ensure that children are meeting their fluid needs (9, 13). For children with impaction, it is advisable to clear stool using osmotic laxatives before increasing fiber intake (12). Children should receive close follow-up to ensure that growth does not slow down and symptoms do not worsen. When feasible, counsel families to support them in moving towards a whole foods, fiber-rich diet. This may involve discussing food availability and access, food preparation skills, and food acceptance.

Dairy Elimination

The role of dairy in pediatric constipation has also been an area of controversy. A growing body of research indicates that for a subsection of children with chronic functional constipation, elimination of dairy leads to resolution of constipation. The recommendation has been controversial in the past because the evidence supporting it was weak; however recently, it has been supported by new research, including high quality, randomized controlled trials with a period of dairy elimination followed by re-challenge (15, 16, 17, 18).

The mechanisms connecting dairy to constipation may vary across patients. There is evidence for an association between cow milk allergy and chronic functional constipation. However, study results have not been consistent, and cow milk allergy is not always present in children who benefit from eliminating dairy. For some patients, the association between cow milk and constipation may be related to mechanisms classified as intolerance or hypersensitivity (15, 16, 19). In some cases, eliminating dairy may be beneficial because a child is consuming an excessive amount of cow milk or because cow milk is satiating and slows motility. Resolution of constipation

may occur with cow milk elimination due to an overall change in diet, as the diet pattern starts to include more water, fruits, and vegetables (15, 18).
Given the promising evidence, it is advisable to trial a 2-4 week dairy elimination for a child with chronic functional constipation that has been resistant to treatment with laxatives and other diet interventions (16). For a child who is exclusively breast/chestfeeding, this would mean having the lactating parent eliminate dairy proteins from their diet. For some children, dairy elimination will be a big sacrifice, while it will be less troublesome for others. It is important to ensure a source of nutrients to substitute for the cow milk or cow milk formula. This is accomplished with hydrolyzed formula or soy milk formula for infants and enterally-fed children. Children older than one year consuming regular foods can receive nutrients they previously received from cow milk by switching to calcium- and vitamin D-fortified soy milk or another fortified non-dairy milk. Soy milk and Ripple pea protein milk have similar protein and calorie content to dairy milk, while most other non-dairy milks contain less. If a child eliminates dairy, counsel the family to ensure adequate sources of protein, calcium, vitamin D, calories and hydration.

Many patients develop a tolerance to cow milk after one year of elimination, and those who do not will have a greater chance of tolerance after more time on the elimination diet. Most children will tolerate cow milk after five years of elimination. Reaction times will vary across children; symptoms may be very delayed, even by as much as two weeks, so it is important to monitor the child re-challenging dairy and have the family keep a food and symptom log to avoid missing the association (15, 16).

Probiotics

Probiotics have been defined as “live microorganisms that, when administered in adequate amounts, confer a health benefit to the host” (20). In recent years, interest in probiotics has been growing, and many families use over the counter formulations on their own initiative to address constipation in their children. Probiotics are a promising avenue for exploration of treatment for childhood constipation, but current evidence does not support a recommendation.

The research on the use of probiotics to treat pediatric constipation shows mixed results, with insufficient evidence from large, randomized controlled trials. Most high quality research in this area is focused on adults, and among trials with children, many studies and reviews have concluded that probiotics did not affect stooling significantly (21, 22). With that said, adverse events have been rare and not serious (21). Research has been inconclusive in part because of differences in study designs, notably in terms of type of organism and dose. Given these differences, many reviews note that it is difficult to evaluate the research as a whole and to build recommendations (20, 22).

There are many proposed mechanisms by which probiotics may influence constipation (20, 21, 23). Dysbiosis, or an imbalance in the microbiome, has been noted in chronic constipation, though

it is not clear if this is a cause or a consequence. This connection has fueled interest in probiotics, but it is not clear what effect modulating the microbiome with probiotics has on stooling. The proposed mechanisms by which probiotics may reduce constipation include increasing release of short-chain fatty acids (which lower intestinal pH and stimulate peristalsis), stimulating water and electrolyte secretion, and promoting anti-inflammatory effects in the mucosa. Much of the research supporting these mechanisms comes from animal studies, and more research is needed to establish clinical results for children (20, 21, 23).

It is important to note that the mechanisms and potential benefits of probiotics are strain-dependent. Additionally, survival inside the body is a key requirement for probiotics' efficacy, and this survival is host- and product- specific. In the United States, probiotics are classified as dietary supplements and are not regulated as closely as medications and food. The Food and Drug Administration does not do any formal testing of probiotics. It can be difficult to know whether the organisms in a probiotic product are the ones specified on the label, whether they are alive, and what will happen to them once inside the body (24, 25, 26, 27). It is particularly important to exercise caution with preterm infants and to assess the quality of the probiotic for this patient population (24).

Ultimately, there is not enough data to recommend probiotics for treating childhood constipation, particularly with regard to specific strains and dosing. However, they are generally safe to take, and there are plausible mechanisms by which they may work.

Table 6-2: Laxatives

Follow instructions on packages, review contraindications, and contact pediatrician if these are not available or if plan needs to be customized. Do not use any product if child has an allergy or sensitivity to any component. Most laxatives should not be used with GI obstruction. Table compiled from Lexi-Drugs (28).

Cause	Onset of Action (hours)	Site of Action	Action	Brand/ Common Name	Comments
Osmotic					
Glycerin suppository	15-30 minutes	Colon	Local irritation; hyperosmotic action	Glycerin	
Lactitol	unknown	Small intestine and colon	Produces laxative effect in colon by causing water influx into small intestine		
Lactulose	24-48 hours	Colon	Delivers osmotically active molecules to colon	Lactulose Cephulac® Chronulac®	Recommended as first-line treatment when PEG not available.
Polyethylene glycol 3350 (PEG)	48 hours	Small and large intestine	Non-absorbable solution that acts as an osmotic agent, causes stool to retain water	Glycolax® Miralax®	Recommended as first-line treatment (1, 2). Dosing: start at 0.4 g/kg/ day and adjust for response. Continue for two months or at least one month after symptoms are gone. For disimpaction, may use 1-1.5 g/kg/day for 3-6 days (2).
Sorbitol 70%	24 to 48 hours	Colon	Delivers osmotically active molecules to colon		Large volumes may result in fluid and electrolyte imbalance.
Saline					
Magnesium Citrate Magnesium Hydroxide	30 minutes to 3 hours	Small and large intestine	Attracts/retains water in intestinal lumen, increasing intraluminal pressure; cholecystokinin release	Milk of magnesia (MOM)	May alter fluid and electrolyte balance.
Sodium Phosphate	2 to 15 minutes	Colon		Fleets enema®	

continued...

Cause (continued)	Onset of Action (hours)	Site of Action	Action	Brand/ Common Name	Comments
Irritant/Stimulant					
Senna	6-10 hours	Colon	Direct action on intestinal mucosa; stimulates myenteric plexus; alters water and electrolyte secretion.	Senokot® Ex-Lax® Fletcher's Castoria® Dulcolax®	Senna recommended as second-line treatment in addition to PEG. Not recommended to add enemas to PEG (2). Enemas may be used for 3-6 days for disimpaction if PEG not available (2). Do not give Biscodyl tablets within 1 hour of antacids or milk products
Bisacodyl Tablets, suppositories, enema	Tablets: 6 to 12 hours Suppository: 15 minutes to 1 hour. Enema: 5 to 20 minutes	Small intestine			
Bulk-Producing					
Methylcellulose Psyllium Wheat dextrin	12-24 hours Up to 72 hours 24 to 48 hours	Small and large intestine	Holds water in stool; mechanical distention.	Citrucel® Hydrocil® Metamucil® Malt Supex® Benefiber®	Should be taken with at least 8 oz fluid. Use of psyllium to normalize stool is backed by clinical research.
Lubricant					
Mineral oil	6-8 hours	Colon	Lubricates intestine; retards colonic absorption of fecal water; softens stool		Contraindicated in infants less than one year or if risk of aspiration. Administer on empty stomach.
Surfactants/Stool Softener					
Docusate/Senna Docusate sodium Docusate calcium	8-12 hours 24-72 hours 24-72 hours	Small and large intestine	Senna- mild irritant; Docusate - stool softener. Detergent activity; facilitates admixture of fat and water to soften stool.	Peri-Colace® Colace® Doss® Surfak®	Do not use Docusate sodium if mineral oil given. Beneficial when stool is hard or dry, or when passage of firm stool is painful.

Table 6-3: Nutrition Interventions For Constipation (1, 2, 3)

Assessment	Intervention	Monitoring/ Evaluation
Clinical/Medical History		
Obtain medical history. Determine primary diagnosis and whether it may affect stooling. Work with PCP or GI specialist to rule out organic disease. PCP may obtain additional tests, including abdominal x-ray, barium enema, intestinal biopsy, rectal or colonic manometry, colonoscopy, or lab tests to identify celiac disease, hypothyroid, hypercalcemia (1, 2).	Provide appropriate care for underlying medical condition in coordination with medical provider. Use laxatives, stool softeners, suppositories, and enemas with care and attention to diagnosis. Polyethylene glycol (PEG) is recommended as first-line treatment, or lactulose if PEG is not available. Milk of magnesia and stimulant laxatives are recommended as additional second line treatment (2). If stool is impacted, medical treatment may begin with a disimpaction regimen, followed by maintenance regimen to prevent recurrence (1, 2, 3). In cases of severe impaction, it may be necessary for patient to be admitted for inpatient disimpaction (1). See Table 6-1 for more information.	Medical contributors to constipation are controlled. When constipation is caused by a correctable factor, re-evaluate need for medications regularly. While medications are being used, work with PCP and patient to adjust dose until two stools per day, or decrease dose if stools are loose. Goal is to produce soft, painless stools and prevent recurrence of constipation. Continue maintenance dose for at least two months, or one month beyond resolution of symptoms. Decrease gradually. Some children need medications indefinitely (2).
Obtain stool history. Determine: • Stool frequency and consistency (Bristol stool chart) • Presence of pain or bleeding • Problem is acute or chronic • Age of onset (if constipation before one month of age or if meconium delayed over 48 hours, may be suspicion for Hirschsprung's Disease) • Toileting techniques, routines • Toilet training history • Known constipation triggers • Frequent triggers for functional constipation, especially changes to routine (4,6) • Presence of impaction, encopresis, or withholding • Presence of abdominal pain, nausea, vomiting, or distention • Previous and current treatment (1, 2)	Address these issues in coordination with medical provider, behavioral therapist, or occupational therapist. Ensure that relevant details are shared with each practitioner. Work with family to create a toileting regimen including regular toilet sits. May recommend footstool to support legs and increase intra-abdominal pressure (1). Include physical or occupational therapist if needed to assist with positioning or special equipment. Parent education about appropriate toilet training methods. Should not be a struggle. Encourage praise or reward for stooling but not punishment (1).	Regular toileting is established (1). Re-evaluate frequently until regular bowel movements are occurring every 1-2 days, with a normal consistency.

continued...

Assessment (continued)	Intervention	Monitoring/ Evaluation
Dietary		
Obtain nutrition history. Assess fluid and fiber intake. Use food record or food recall, along with interview about dietary preferences and practices. Assess: - Energy adequacy - Balance of macronutrients - Sources of soluble and insoluble fiber - whole grains, fruits, vegetables, legumes, nuts, and fiber-enriched products. - Estimated fiber needs (DRI goal of 14 g/1000 kcal or AAP goal of 0.5 g/kg/day to maximum of 35 g/day, or age in years + 5g/day to a maximum of 10g/day) (8, 10, 14) - Hydration needs and status - Specific foods that trigger constipation episodes Recent changes to intake such as initiation of solids or cow milk in infants, or new dietary patterns in older children and teens (1, 2)	Provide education and counseling to support dietary changes. Increase fluids as needed, especially water. Consider including one serving per day of peach, pear or prune juice, or other sorbitol-containing juice. Evidence does not support providing extra fluid beyond estimated needs, but it's important to ensure child is meeting needs (2). Increase foods with high water content such as fruits and vegetables. Increase dietary fiber as needed by incorporating whole foods into diet: • Whole grains such as brown rice, whole wheat bread, etc. • Raw, cooked, or dried fruits • Raw or cooked vegetables • Legumes (beans, peas, lentils) • Nuts and seeds Provide practical tips for adding fiber-rich foods to diet, such as: • Using nuts/seeds in baking • Mixing white and brown rice if brown alone is rejected • Offering chopped vegetables and fruit slices with dip as a snack • Adding beans, lentils and vegetables to pasta sauces, soups, chilis and casseroles • Making custom trail mix with dried fruit, shredded coconut, and nuts May consider 2-4 week trial of cow milk-free diet. Use supplemental fiber products with caution. Coarse wheat bran and psyllium are supported by clinical evidence (11, 12). For enterally-fed children, may increase water or add sorbitol-containing juice. Can also try home-blended tube feedings made from real food; commercial blended food tube feeding products such as Nourish, Pediasure Harvest®, or Compleat Pediatric Organic Blends®; or standard formulas with fiber, such as Nutren Jr. with Fiber®, Enteral Pediasure with Fiber®, Peptamen Jr. with Fiber®, Promote with Fiber®, or HN Fibersource®.	Intake of fluid and fiber is improved. Re-evaluate diet frequently until stools are 1-2 per day with normal consistency. Be on the lookout for key transition points often associated with constipation, such as introduction of solids and introduction of cow milk. Provide anticipatory guidance, including dietary counseling, at these key transition points (4, 6).

Dietary (continued)		
Assess level of physical activity.	Increase physical activity as tolerated.	Recommended activity schedule is followed.
Psychosocial		
Assess need for behavioral intervention.	Connect family to psychologist or community resources to address behavioral challenges as needed.	Follow up as needed.

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Chapter 7

NUTRITION INTERVENTIONS FOR DIARRHEA

Naomi Katsh, MD
Updated by Khimberly Schoenacker, RD, CSP, CD

Diarrhea is the sudden increase in frequency and looseness of stools. Diarrhea is defined by the World Health Organization as the passage of three or more watery stools in 24 hours or three times the normal number of stools in 24 hours (1,2,3). The best indicator of the severity of diarrhea is the frequency. If severe or chronic, this condition has a high potential for morbidity (and mortality, especially in developing countries). Decreased morbidity and mortality have occurred because of improved hydration status.

The main complication of diarrhea is dehydration from the loss of fluid and electrolytes with the stools. Nutritional complications may also develop, especially in chronic situations. Depending on the severity, chronicity, and underlying medical condition, evaluation and treatment of diarrhea may require input from many different health professionals including nurses, registered dietitian nutritionists (RDNs), primary care providers, and gastroenterologists.

Although there is not always a clear distinction, the subject of diarrheal illness can be approached as either an acute or chronic problem.

Acute Diarrhea

Acute diarrhea refers to a self-limited illness usually of less than 2-3 weeks duration.
Acute diarrhea may lead to electrolyte imbalance and dehydration, which can be life threatening. Infants and young children with diarrhea are more susceptible to dehydration than older children and adults because of their smaller intravascular
volume and a lower capacity to concentrate urine (2). Children and youth who have disabilities that affect oral-motor function are already at risk for dehydration due to impaired lip closure or swallow and may be at even greater risk because of difficulty replacing the fluids lost through diarrhea.

Causes

There are many causes of acute diarrhea. Some common causes include:

- Infection: Diarrhea in children is usually caused by a viral infection of the lining of the intestine (gastroenteritis) and can be accompanied by vomiting, fever, and abdominal pain. Types of

infection include:

1. Viral: (e.g., rotavirus, enteric adenovirus). In the United States, viruses account for at least 30-40% of episodes of acute gastroenteritis.
2. Parasitic: (e.g., Giardia)
3. Bacterial: (e.g., E. coli O157, Salmonella, Shigella, Campylobacter)
4. Non-gastrointestinal infection: Diarrhea can accompany otitis, pneumonia or urinary tract infection

- Medication side-effect (frequent with some antibiotics)
- Food intolerance

Nutritional Complications

Nutritional complications from acute diarrhea may not always be evident. Nutrient deficits are uncommon among previously healthy children with self-limited gastroenteritis. Dehydration is the most concerning complication of acute diarrhea. In the literature, there are multiple definitions of the degrees of dehydration. Table 7-1 describes one approach to the assessment of dehydration for those with acute diarrhea.

Table 7-1: Assessment of Dehydration (1,2,4,5)

	Percent Body Water Lost	Signs and Symptoms
Minimal dehydration	1 to 2%, subclinical	Increased thirst and mild oliguria (decreased urine output)
Mild dehydration	3 to 5%	Increased thirst, oliguria, mucous membranes slightly dry
Moderate dehydration	6 to 9%	Marked thirst, urine output <1mL/kg/hr, dry mucous membranes, decreased or absent tears, slightly sunken fontanel, sunken eyes, delayed capillary refill, may have increased heart rate, may be listless and/or irritable
Severe dehydration	>9%	All the signs of moderate dehydration and may have hypotension; weak pulse; bradycardia or tachycardia; cool, cyanotic extremities; severe lethargy; recoil on skin turgor test in less than 2 seconds

After rehydration is started, refeeding the intestinal tract is recommended as the appropriate dietary management (1-3,4,6,7). There are multiple physiologic effects when the gastrointestinal tract receives no enteral nutrition. Starvation has been shown to cause atrophy of the gastrointestinal mucosa, decreased production of digestive enzymes, and increased permeability of the mucosal barrier. All of these effects can lead to decreased ability of the gut to absorb nutrients (6,7).

Some children and youth with special health care needs may experience nutrition deficits from repeated bouts of mild acute diarrhea, even when appropriately managed.

Treatment

Mild acute diarrhea requires no special treatment. Adequate fluid intake should be a priority, but a strict clear liquid diet is no longer the treatment of choice. Data indicates that feeding with the usual diet is appropriate for most cases of acute diarrhea (1-8). If an infant is breast/chestfed, this should be continued on demand and other fluids given if needed for supplementation. Breast/chestmilk contains substances which may stimulate and protect the gastrointestinal mucosa (6). Formula-fed infants also should be continued on their routine formula. Milk and milk-based formulas have historically been avoided during episodes of diarrhea.
There may be mucosal damage during the illness that creates temporary lactase deficiency. However, at least 80% of children do not have worsening diarrhea from this temporary lactase deficiency and can safely consume milk-based formula or milk (6,7). Change to a lactose-free formula is only recommended if stool output increases on a milk-based formula (3).

Previous recommendations for treatment of acute diarrhea were for a period of "bowel rest" with clear liquids only, then a gradual re-introduction of first diluted, then full-strength formula or milk along with a very limited diet of solid foods. Diluted formula is no longer recommended. Current information has demonstrated that early feeding of a routine diet leads to a better overall outcome. Specific beneficial effects are decreased duration of illness, improved weight gain, and improved nutritional state (4,6,7).

What constitutes the most appropriate mixed diet for feeding during acute diarrhea is discussed in the medical literature. Historically, the BRAT diet (banana, rice, applesauce and toast/tea) was recommended (9). This very restricted diet is high in carbohydrates but very low in calories, fat, fiber, protein, calcium, and Vitamins A and B12 (7). Current, broad guidelines for an appropriate mixed diet are for food that is palatable, inexpensive, culturally acceptable, and easily digested (4,6). This may include complex carbohydrates (rice, wheat, potatoes, bread, cereal), lean meats/poultry, eggs, fruit, vegetables, and yogurt (4).

Medications are generally not prescribed or recommended for infants or children with acute diarrhea. Viruses are the predominant cause of acute diarrhea, especially in developed countries; therefore, antibiotics are not indicated when treating acute diarrhea illnesses. Very few studies evaluating medications have been done with children to demonstrate safety or efficacy. The potential risks of medications, including antispasmodics and antimotility agents, outweigh any potential benefits (1,2,4,6).

Mild (3-5%) to moderate (6-9%) dehydration resulting from acute diarrhea can be treated with oral rehydration. There are commercially available preparations (e.g. Pedialyte®, Rehydralyte®) for oral

rehydration. In developing countries, child mortality from diarrheal illness, while still unacceptably high, has decreased dramatically due to oral rehydration solution (ORS) programs for treatment and prevention of dehydration.

Since 1975, The World Health Organization (WHO) and the United Nations Children's Fund (UNICEF) have recommended the formulation of ORS used in developing countries. . Various modifications to the standard ORS have been derived (10). These modifications have included hypo-osmolar or hyperosmolar solutions, use of rice-based ORS, zinc supplementation, and the use of amino acids, including glycine, alanine, and glutamine. Some of these variations have been successful, some have not, and others are still under investigation. Lower osmolarity ORS has been found to be more effective for acute, non-cholera diarrhea in children and causes fewer cases of hypernatremia (high sodium).

Osmolality is a major factor in determining the efficacy of ORS. In general, juice, broth, carbonated beverages, and sports drinks should not be used for oral rehydration because their high osmolalities may induce osmotic diarrhea, and the electrolyte content is not appropriate (1-3,4,6,7). Diluted juice, broth, and sports drinks can be used for some children if other, more appropriate rehydration fluids are not available. Cereal-based oral rehydration therapy has also been proposed as a method of rehydration, which also provides some nutrients (6,11). There are no commercially available, cereal-based products at the time of this writing. See Resources at the end of this chapter for a recipe for cereal-based ORT solution.

Chronic Diarrhea

Diarrhea is considered to be chronic if one episode lasts longer than three weeks or if there are multiple episodes with only a few weeks or months between.

Causes

Some of the same factors that cause acute diarrhea may also result in chronic diarrhea. In addition, there are other etiologies of chronic diarrhea. Some of the more common ones include:

- Carbohydrate intolerance (e.g., lactose, fructose)
- Other food/formula intolerances, food allergies, improper formula preparation, tube-feeding complications
- Chronic, nonspecific diarrhea (This is a term used for diarrhea of at least 3 weeks duration, greater than 3 loose stools per day, and no evidence of malabsorption or enteric infection.)
- Cystic fibrosis (see Chapter 16)
- Celiac disease (Gluten-sensitive enteropathy)
- Short bowel syndrome (see Chapter 18)
- Inflammatory bowel disease (Crohn's disease and ulcerative colitis)
- HIV/AIDS and other immune deficiencies

- Constipation/obstipation with encopresis
- Pseudomembranous colitis (Most often related to antibiotic use)
- Micronutrient deficiency (e.g., zinc deficiency can be both a cause and a complication of chronic diarrhea (7))

Nutritional Complications

Compromise of nutritional status is much more likely to occur with chronic diarrhea than with acute diarrhea. Malnutrition can result both from chronic loss of nutrients and fluid through the gastrointestinal tract and from overzealous attempts at dietary eliminations to determine the cause of chronic diarrhea (7). In turn, this malnutrition can lead to additional diarrhea secondary to alteration of mucosal absorptive ability and decreased enzyme activity (7). Children who have chronic diarrhea may have decreased appetites and therefore, decreased intakes of nutrients.

Treatment

Treatment of chronic diarrhea depends on the cause of the diarrhea and the results of a total assessment. Malnourished infants with diarrhea present a significant challenge for successful treatment and need energy replacement in addition to rehydration. Energy requirements of infants or children with chronic diarrhea may be as high as 200 kcal/kg/day (2). Enteral feedings may be attempted orally or by slow continuous nasogastric tube feeds. For children who are severely malnourished or who have poor gastrointestinal function due to other causes (e.g., short bowel
syndrome), parenteral nutrition may be required. Medications may have some role in treating chronic diarrhea; however, one should never give over-the-counter antidiarrheal medication to a child without first consulting with their pediatrician. Pancreatic enzyme replacement is required in cystic fibrosis and other pancreatic disorders. Sulfasalazine and corticosteroids may be used in inflammatory bowel disease. If there is a specific protein or carbohydrate intolerance or enzyme deficiency, avoidance of the offending foods is the treatment of choice.
Many children affected by chronic diarrheal conditions may require nutrition evaluations and follow-up throughout infancy and childhood. Special formulas and dietary supplements may be needed. Probiotic and prebiotic supplementation may assist with treatment and recovery, though current research does not provide specific dosage recommendations.

Diarrhea is a very common occurrence in childhood. Frequency and duration of the stools are two variables used to determine what, if any, evaluation is needed. The remainder of this section presents guidelines for nutrition assessment, intervention, and evaluation/outcome for children and youth with acute (Table 7-2) and chronic (Table 7-3) diarrhea.

Table 7-2: Nutrition Interventions for Acute Diarrhea

Assessment	Intervention	Evaluation/Outcome
Anthropometric*		
Measure and plot on appropriate growth chart: • Height or length for age • Weight for age • Weight for height (or length) or BMI • Head circumference (under age 2 years) Compare current measurements to available previous measurements. If there are recent weights, this can be helpful in assessing amount of dehydration. Repeat height/length, weight and head circumference (OFC) measurements at every clinic visit.	Adjust recommendations for energy intake based on growth data.	Child maintains growth pattern.
Clinical		
Obtain information about clinical history. Include child's age, other diagnoses (prematurity, congenital disorders, prior surgery), medications, and possible exposures through day care attendance, camping, or foreign travel. Obtain information about stool history, including duration of illness, stool frequency and volume, consistency, and presence of blood or mucus.	Attempt oral rehydration to prevent need for hospitalization and parenteral fluids. Oral rehydration therapy with a glucose- electrolyte solution is only required if there is dehydration. 50-100 ml/kg of body weight of solution is given over a 4-hour period. Ongoing losses can be estimated at 10 ml/kg for each stool (4).	Patient is rehydrated; hospitalization and administration of parenteral fluids are avoided.
Work with primary care provider or medical consultant. Medical evaluation may include some of the following when indicated: • Physical examination, especially assessment of hydration status (see Table 7-1 for clinical description of degrees of dehydration) • If diarrhea is very frequent, prolonged or bloody, tests may be indicated, including stool culture for bacteria, tests for rotavirus or parasites, and stool white blood cell tests. • Blood tests for electrolytes may be done, especially if hospitalization is required.	Treatment depends on the cause of diarrhea. Provide medical management and appropriate nutrition intervention for diagnosed diseases/ disorders. Generally, antidiarrheal medications are not recommended.	Appropriate medical and dietary recommendations are followed. Physical signs of dehydration should resolve, and diarrhea should gradually decrease in severity. If diarrhea does not resolve, further medical testing and management may be indicated.

Dietary		
Obtain a diet history and compare with stool history to determine possible relationships between foods and diarrhea. If further information is needed, request a 3- to 7-day food record and a 3- to 7-day stool record.	Dietary triggers of diarrhea are identified and eliminated from food pattern. Use prepared glucose electrolyte solutions for rehydration if needed. High carbohydrate drinks are inappropriate. Rapid refeeding of usual diet is recommended.	Diarrhea and dehydration are resolved. Food pattern provides adequate amounts of energy, protein, and vitamins and minerals.

*See Chapter 2

Table 7-3: Nutrition Interventions for Chronic Diarrhea

Assessment	Intervention	Evaluation/Outcome
Anthropometric*		
Measure and plot on appropriate growth chart: • Height or length for age • Weight for age • Weight for height (or length) or BMI • Head circumference (under age 2 years) • Mid-upper arm circumference Conduct Pediatric Nutrition Focused Physical Exam Calculate rate of weight gain and linear and head circumference (OFC) growth. It is very important to know if there has been weight loss or slowing of weight gain. Repeat height/length, weight and OFC measurements at every clinic visit.	Adjust recommendations for energy intake based on growth data and risk for malnutrition.	Child maintains growth pattern.
Clinical		
Obtain information about clinical history. Include child's age, other diagnoses (prematurity, congenital disorders, prior surgery), medications, possible exposures through day care attendance, camping, or foreign travel. Obtain information about stool history, including duration of illness, stool frequency, volume, consistency, and presence of blood or mucus. Specific emphasis should be placed on the characteristics of the stool and if there is a family history of gastrointestinal disorders. A careful review of systems must be done to see if other body systems are involved (e.g., respiratory tract symptoms might be the clue that cystic fibrosis is the cause of chronic diarrhea).	Treatment of chronic diarrhea is dependent on the cause. Medical and dietary management is available for many of the disorders that cause chronic diarrhea.	Treatment for medical causes of chronic diarrhea alleviates symptoms.

Work with primary care provider or medical consultant. Medical evaluation may include different studies, depending on the clinical situation, diagnosis (if known), and duration and severity of diarrhea: • Stool cultures and studies may be done for infectious causes and fat content • Blood tests may be done for electrolytes, specific micronutrients: vitamin E and B12 (if problems with ileal absorption), total protein, albumin, d-xylose, carotene • Sweat test • Gastrointestinal x-rays and/or sigmoidoscopy or colonoscopy and biopsy Physical examination to include not only assessment of hydration status, but also assessment of nutritional status.	Medication may be indicated in some cases (e.g., pancreatic enzymes in cystic fibrosis, sulfasalazine or corticosteroids in inflammatory bowel disease).	Close, frequent follow- up is indicated to see if appropriate medical recommendations are being followed or are effective in decreasing the amount or frequency of diarrhea.
Dietary		
Obtain a diet history and compare with stool history to determine possible relationships between foods and diarrhea. For example, evaluate whether or not onset of diarrhea coincides with introduction of cow's milk or cow's milk protein formula (cow's milk protein sensitivity, lactase deficiency); cereals or bread (gluten- sensitive enteropathy); foods with table sugar added (sucrase deficiency). Consider obtaining a 3-7day food record and a 3-7day stool record.	If food allergy or intolerance is suspected, try eliminating specific foods that seem to be related to the diarrhea. Caution must be taken that an elimination diet is not so extreme that it leads to inadequate nutrient intake. Provide instruction about special diets when indicated (such as lactose-free diet for lactase deficiency, gluten-free diet for gluten-sensitive enteropathy, low fructose for fructose intolerance, low FODMAP diet, etc.) (5-7).	Close, frequent follow-up is indicated to see if appropriate dietary recommendations are being followed or are effective in decreasing the amount or frequency of diarrhea. Food pattern provides adequate amounts of energy, protein, and vitamins and minerals.
Evaluate ratio of energy from fat and carbohydrate in the diet (low fat diet may contribute to nonspecific diarrhea) (3,8). Evaluate volume of liquids ingested and amount of fruit juice consumed (3,8).	For chronic nonspecific diarrhea, consider decreased fruit juice intake and increased fat and fiber intake (8,12).	Food pattern does not contribute to diarrhea.
If child is tube-fed, evaluate the type and preparation of formula, rate of feeding, tube position (gastric or small bowel), care of feeding bags and tubes, etc. Consider changing to a formula with added fiber (3,12).	Consider adjustments to tube feeding formula, rate, as indicated.	Tube feeding does not contribute to diarrhea.

*See Chapter 2

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Resources

Recipe for cereal-based ORT (Oral Rehydration Therapy) solution (11)

½ - 1 cup dry infant rice cereal
2 cups (16 oz) water
¼ teaspoon table salt

Measure salt and dissolve in water. Gradually add cereal to the water until the mixture is as thick as is drinkable. Mix well. Discard after 6 to 8 hours or if it becomes too thick to drink.

Chapter 8

ORAL-MOTOR AND SWALLOWING PROBLEMS

Robin P. Glass, MS, OTR/L and Lynn Wolf, MOT, OTR/L

The diagnostic criteria for Pediatric Feeding Disorder include a dysfunction in feeding skills leading to:

- Need for texture modification of liquid or food
- Use of modified feeding position or equipment
- Use of modified feeding strategies

Oral-motor and swallowing problems often underlie feeding skill dysfunction and can limit a child's ability to consume adequate nutrition in a variety of ways. The child may have inefficient or abnormal oral-motor patterns, making ingestion slow or labored. Oral-motor difficulties may limit the types of foods the child can eat. Swallowing dysfunction may severely restrict the type of food textures that are safe for the child to eat. Most frequently, swallowing dysfunction affects the child's ability to drink liquids. This may not only compromise the child's overall nutrition but also their hydration status. Some factors associated with oral-motor and swallowing problems include:

- Abnormalities in muscle tone and delays in motor development
- Oral-facial defects that interfere with feeding (such as cleft palate)
- Anatomic defects in the airway that compromise breathing
- Delayed oral motor development or abnormal oral motor patterns (e.g., a tonic bite reflex)
- Hypersensitive responses to touch, taste, and/or temperature in and around the mouth
- Dental problems, such as severe dental cavities or malalignment of the jaws and teeth
- Inability to properly coordinate feeding, swallowing, and breathing due to chronic medical conditions or central nervous system damage
- Related medical conditions, such as gastroesophageal reflux or constipation that may affect willingness to eat

Oral-motor and swallowing problems can lead to prolonged length of feeding sessions and disruption of the feeding relationship. The child and family can experience emotional distress and can become physically depleted from the amount of energy expended in the feeding process. If feeding difficulties such as these are observed, a thorough feeding evaluation is indicated. A pediatric occupational, speech, or physical therapist skilled in feeding disorders can perform this type of evaluation.

Complete evaluation of oral-motor and swallowing deficits in children with feeding challenges

involves assessment of a wide range of factors beyond the level of oral-motor control. These include assessment of:

- Basic postural control and level of motor development as it relates to feeding
- Tactile sensitivity in the oral area and throughout the body
- The child's ability to achieve and maintain appropriate behavior and state of alertness for feeding
- The stability of the child's physiologic functions, such as heart rate or respiratory rate, to support feeding
- Coordination of feeding, swallowing, and breathing
- Swallowing function evaluated by clinical observations and referral for videoflouroscopic swallowing study (VFSS), if indicated

In addition, difficulties in parent-child interaction or mealtime management may also be associated with oral-motor feeding problems. It is often difficult to differentiate between physical, behavioral, and interactional problems. Thus, it is necessary to consider all three when evaluating a child for oral-motor feeding problems (1).

Related medical problems, such as gastroesophageal reflux (GER), may also influence the child's feeding ability. If the child associates eating with the pain that often accompanies GER, the child may be resistant to feeding, and food intake may decline dramatically. If GER is suspected, further evaluation by the child's primary care physician or a gastroenterologist is indicated.

Most feeding problems have multiple underlying factors that contribute to the overall nutritional and feeding deficits. For this reason, a team approach is the most effective method to adequately assess and intervene with the oral-motor feeding problems. In addition to the occupational, speech, or physical therapist, this team should include a registered dietitian nutritionist (RDN), a pediatrician (or other primary healthcare provider), and the caregivers. The team may also include a nurse, social worker, or other mental health provider to address the psychosocial issues that frequently occur concurrently with the feeding problem (see Chapter 9).

Appropriate interventions can significantly improve the child's basic feeding skills as well as improve the ease of feeding for the caregiver. Mealtimes can become more satisfying for the child and family, promoting better growth and nutrition. Intervention may include:

- Proper positioning during feeding
- Specific therapeutic activities to improve basic oral-motor skills
- Specific food types or textures to support the child's level of oral-motor control
- Dietary modifications to support swallowing function
- Adapted feeding utensils
- Specialized feeding techniques

If the child is still unable to consume an adequate intake and/or it is unsafe for the child to eat, partial or full nutrition may need to be given via a nasogastric or gastrostomy tube (see Chapter 10).

Planning and implementation of any feeding interventions require collaboration with the caregivers and the other members of the team. Treatment strategies should be arranged so they support caregiver priorities, improve nutrition, and address underlying oral-motor, swallowing, and feeding problems.

Table 8-1 describes the developmental sequence of oral-motor and self-feeding skills. Table 8-2 presents guidelines for nutrition assessment, intervention and evaluation/outcome for children with oral-motor feeding problems.

Table 8-1: Developmental Sequence of Oral-Motor and Self-Feeding Skills

Age	Reflexes	Jaws and Cheeks	Lips	Tongue	Swallowing	Self-Feeding
Term-2 months Breast/ Bottle Feeding	• Sucking • Palmomental • Rooting • Gag • Phasic bite	• Fat pad present • Primary jaw movement downward during sucking	• Closed at rest • Form a seal around breast or bottle	• Fills oral cavity • Is inside mouth behind lips • Creates pressure forces during sucking: compression and suction	• Suck-swallow sequence 1-3:1 • Occasional cough or choke; esp with high flow • Air swallow common	• Hand-to- mouth activity • Expects feeding at regular intervals
2-4 months Breast/ Bottle Feeding	• Voluntary sucking emerges; reflexive sucking integrated • Can voluntarily inhibit suck to look or listen	• Buccal cavity begins to develop	• Smacks lips	• Tongue protrudes in anticipation of feeding or if nipple touches lip	• Air swallow common • Infrequent coughing or choking during feeding • Suck-swallow sequence remains 1-3:1	• Brings hands and toys to mouth • May gag on fingers/toys • Pats bottle or breast
4-6 months Starting pureed foods	• Oral reflexes integrated into voluntary skills	• Able to grade jaw movement during sucking • Opens mouth for spoon	• Closes mouth/lips around spoon • After practice with puree, draws in lower lip when spoon removed	• Tongue protrusion pattern may push purees out of mouth • Tongue quiet/still as mouth opens for spoon • Tongue protrudes at moment swallow is initiated	• Choking rare on breast or bottle • Safely swallows pureed foods • More than one swallow per bite	• Purees may be introduced • Reaches for or plays with spoon • May gag with new foods
6-8 months: Adding textured spoon foods and finger foods	• Oral reflexes integrated into voluntary oral-motor control	• Chewing: Up and down munching and biting • Inner cheeks draw inward during eating • Jaw closes on solid then sucks it • Jaw held closed while a piece of soft solid is broken off	• Blows “raspberries” • Upper lip moves downward and forward to actively clean spoon	• Tongue begins lateral shift when food is at side of mouth	• Able to swallow mashed and meltable foods • Can choke on more textured foods	• Begins finger feeding • Feeds self cracker • May hold bottle • Inserts spoon crudely into mouth

8-12 months Adding cup drinking	• Voluntary oral control	• Chewing: Munches with diagonal movements as food is transferred from center to sides • Uses cheeks to stabilize food on chewing surface • Voluntary biting on • food and objects	• Lips active with jaw during chewing • Closes lips on cup rim	• Lateral movements to transfer food from center to sides of mouth • Lateralizes from one side across midline to other side	• Cup may be introduced; expect coughing/ choking • One sip at a time from an open cup • Once comfortable with cup, takes 1-3 sucks before stopping to swallow and breathe	• Precise finger feeding • Better precision with spoon feeding • Can self-feed a full meal
12+ months		• Controlled, sustained bite on soft cookie • Begins rotary chewing movements	• Lips closed during swallow with no food or liquid loss • Lower lip is drawn inward to be cleaned by upper gums	• Licks food from lower lip • Intermittent tongue tip elevation	• May take all liquids by cup • Takes 4-5 continuous swallows from cup • Swallows ground, mashed or chopped table foods without gagging	• Independent drinking from cup with lid • Manages open cup with supervision • Spoon feeds with control

Table 8-2: Nutrition Interventions for Oral-Motor Feeding Problems

Assessment	Intervention	Evaluation/Outcome
Inability to ingest age-appropriate food textures and liquids Inability to take age-appropriate amount of food/ liquids for adequate growth in a reasonable length of time (see Chapter 1) Presence of abnormal oral-motor patterns (See Table 8-1 Developmental Sequence of Oral-Motor and Self-Feeding Skills)	If observed, refer to a pediatric occupational, physical, and/or speech therapist for further evaluation and intervention	The feeding therapist evaluates feeding skills and develops an intervention plan. Improvements may be observed in basic oral-motor skills, volume of food ingested, decreased feeding times, and/or safer feeding.
Observe swallowing function: coughing; choking; wet sounding breathing during feeding; difficulty catching breath during feeding; frequent respiratory infections	If observed, refer for a clinical feeding evaluation and possible videofluoroscopic swallowing study (VFSS).	Diet modifications to increase safety or ease of feeding may be made based on results of feeding evaluation or VFSS. If oral feeding is unsafe, tube feeding may be necessary. See Chapter 10.
Observe sensory responses: gagging with food or feeding tools; resistance to touch around face and mouth; excessive selectivity of food tastes/ textures.	If observed, refer to occupational, physical, or speech therapist for an in-depth sensory and sensory integration assessment.	Intervention by occupational, physical, or speech therapist leads to improvements in acceptance of age-appropriate food textures, an increase in volume, and more timely food ingestion
Feeding resistance or refusal is observed. Differentiate underlying cause; consider medical (GER, swallowing or respiratory problem) versus parent/child interaction versus sensory issues.	Interactional: see Chapter 9 Medical: Refer to PCP or specialist, such as gastroenterologist Sensory: Refer to feeding specialist	Referral intervention leads to greater ease of feeding for parent and child.
Observe parent/child dynamics: this includes behavioral interactions; parent expectations; emotional tone of meals; mealtime structure.	If problems are noted with either parent/child interaction or feeding behaviors, see Chapter 9. Referral to a behavior specialist may be indicated.	Referral intervention leads to improvements in parent/child interactions and feeding behaviors.

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Chapter 9

BEHAVIORAL ISSUES RELATED TO FEEDING

Yev Veverka, PhD, BCBA-D
Anna Joseph, MEd, BCBA

Introduction

Meera took her four-year-old son, Ravi, to his annual check up with his pediatrician. In the waiting room, she filled out the forms. "Are you concerned about your child's weight or eating habits?" Meera thought about the ongoing mealtime battles in her home and checked "yes". "Does your child eat a good variety of foods (i.e., meat, vegetables, grains, and fruit)?" Meera checked "no" without hesitation. Ravi mostly snacked on his favorite crackers throughout the day in addition to drinking whole milk from a bottle. He never made the switch to 2% milk, and Ravi refused to drink out of a cup.

When the pediatrician came in, he asked Meera to expand on her answers to the questionnaire. Meera explained that Ravi refuses to sit down for meals and that each mealtime is a battle. When she or her husband offers Ravi a new food, he refuses for it to be placed anywhere near him. He screams and pushes the food away until his parents give up and remove it from the table. Meera reported giving into Ravi's yelling and allowing him to just eat one of his favorite foods.

The pediatrician pulled up Ravi's growth chart and pointed out that Ravi is growing well along his curve. The pediatrician talked about the importance of continuing to offer nutritious foods and how Ravi is demonstrating behavior that is very common at this age. The pediatrician said that they will check in again at his next check up. While Meera feels reassured that Ravi is on track with his growth, she does not know what to do about the mealtime battles. She is still anxious and unsure about how to improve family meals.

Meera and Ravi's story is a common one. Mealtime challenges are highly prevalent in the preschool years. Selective eating is one of the most frequently reported mealtime concerns. Carruth, Ziegler, Gordon, and Barr found that 19% of children were perceived as selective eaters by their parents at age four months, and this number increased to 50% at 24 months (1). Though there is much variation in prevalence estimates across studies, many have found selective eating to be predominantly present during the preschool years (2,3). It is hypothesized that transient selective eating is a part of normal child development. However, selective eating can persist and lead to the development of clinical disorders (2). Because both trajectories present similarly in early childhood, the long term course of selective eating is not clear.

Mealtime challenges can lead to negative health impacts and stress on the family. Rather than waiting to determine if these challenges will resolve with time, a behavioral approach to mealtime and feeding challenges can be useful for identifying, assessing, and treating mealtime challenges. The field of Applied Behavior Analysis (ABA) has provided empirical support for methodology to assess and intervene in order to decrease challenging mealtime behaviors while increasing appropriate mealtime behaviors.

The following is a discussion of a behavioral approach to the assessment and intervention of mealtime challenges. The behavior analyst is responsible for a series of tasks that will be defined and explained in this chapter. These consist of (a) defining a challenging behavior, (b) determining why it is occurring, (c) writing a meaningful objective for behavior change, (d) determining a set of procedures for behavior change, (e) collecting data to measure progress, and (f) providing ongoing progress monitoring throughout the intervention and changing procedures when necessary.

Behavioral Assessment to Identify and Define Behavior

Grace, a Board Certified Behavior Analyst (BCBA), was asked to consult with Ravi's family. Grace started by individually interviewing Ravi's parents about their mealtime concerns. Ravi's mom, Meera reported that she felt guilty about the lack of nutrition that Ravi gets from his diet. She told Grace she feels worried about trying to wean from the milk bottle because she's afraid Ravi would not meet his energy needs. As long as Ravi ate nutritious foods, Meera did not care if she had to cook him separate meals or allow him to watch TV at the table.

Arjun, Ravi's dad, reported feeling stress because Ravi refused to sit with them at mealtimes. He stated that chasing Ravi around with food made it so that he and Meera could never enjoy a meal together anymore.

Grace also gathered information about Ravi's favorite foods, timing of his meals, and who was present during mealtimes. Further, Grace asked questions about the family's culture and what values and goals they had as a family around mealtime. Grace learned that in their culture, children are often fed by the parents and that naan, an Indian bread, is used to scoop food.

We will discuss two main purposes of behavioral assessment: defining the behavior of interest and determining the function of the behavior. First, behavioral assessment is used to define the behavior of interest. In ABA, this is commonly referred to as describing the topography of behavior, or what it looks like. In defining a mealtime challenge, the behavior analyst determines an operational definition for the behavior. An operational definition is one that is observable and measurable. An operational definition focuses on the behavior of the child, or what they are doing rather than how they are feeling. An operational definition is important in order to measure behavior and accurately track progress of behavior change. For example, if one were to say "Ravi

is demonstrating a lot of anxiety during mealtimes," you might imagine a set of behaviors that is very different from one of your colleagues. However, if you heard "Ravi is crying and pushing food away each time a new food is offered," you and everyone else observing Ravi would know exactly what they are looking for and what to count as an occurrence of the challenging behavior.

In order to determine an operational definition, a behavior analyst needs to conduct initial assessments to understand what the behavior(s) of concern are, what they look like, and how often and when they occur. Behavior analysts use both indirect and direct methods of assessment in order to achieve this goal. Direct assessment will be discussed later in this chapter. In an indirect assessment, information is gathered without directly observing the behavior. A common indirect assessment method for understanding mealtime behaviors is a caregiver interview. This allows for the gathering of data based on a caregiver's account of the development of mealtime challenges, previous interventions that have been attempted and their success, description of mealtime challenges, and possible situational variables that maintain the challenges (4). The interview process is also a time to gather information about the child's medical history, medications taken, and g-tube feedings (when applicable), as well as to review reports from related disciplines, such as physicians, registered dietitian nutritionists, and occupational therapists (4, 5).

An important goal in a behavioral approach to intervention is to assess the social significance, or the extent to which intervention will impact the quality of life of the child who is being assessed. Interviewing caregivers is an effective starting point to assess social significance (6). Asking about the effects that various mealtime problems have on the daily routines, caregiver stress levels, and family functioning can provide valuable information about the importance of intervention for each behavior. For example, while a child not sitting for mealtime may be a major problem for a family that strives to sit together at mealtimes, it may not be an issue at all for a family that eats separately on the couch.

A behavior analyst's role consists of determining who to interview, what questions to ask, and what further assessments to conduct based on interview results. After this initial gathering of information through indirect assessment methods, a behavior analyst can determine what behavior needs to be observed directly in order to gather further information about challenging mealtime behaviors (direct assessment).

After Grace gathered information from Ravi's parents, she planned to observe some family mealtimes to examine the behaviors that Ravi's parents mentioned. Grace chose to collect data on the challenges that both Meera and Arjun noted in their interviews: leaving the table or refusing to come to mealtime, food refusal, and food selectivity.

Direct assessment measurement techniques are also important for selecting and defining target mealtime behaviors. Direct observation is guided by the behaviors discussed during indirect

assessment interviews. For example, if a caregiver mentions that a big stressor is their child's leaving the table, the behavior analyst would directly assess this behavior, tracking the number of times or the duration of time the child is seated versus away from the table. If the caregiver is concerned about selective eating, the behavior analyst would track the number and quantity of each food eaten, as well as the occurrence of refusal behaviors.

A common method of direct assessment is the observation of natural mealtimes (4,7,8,9). Ideally, direct observation consists of observing mealtime in the natural environment under normal conditions (4). For instance, in the example of Ravi, Grace might observe a dinner at home, as well as a lunchtime at school. Sometimes, behavior analysts might do a mealtime observation in a clinical setting, in which a variety of foods from different food groups are presented and caregivers are instructed to conduct a meal as they typically would (10).

Direct assessment allows the behavior analyst to determine how to define and measure a behavior and how to take data. It is important to define challenging mealtime behaviors in order to develop appropriate intervention methods. Another benefit is the ability to measure baseline levels of behaviors. This provides a starting point for measuring progress to determine if treatment is effective. Further, direct assessment provides objective information on both caregiver and child behaviors, which may not be identified in caregiver interviews.

Assessment to Determine the Function of a Behavior

Through direct assessment, a behavior analyst is also responsible for determining how a child interacts with their environment. Operationally defining and observing behavior for baseline levels is an appropriate first step in the identification and definition of challenging behaviors. However, simply defining the behavior is not sufficient in developing a behavioral intervention plan. An important component is looking for patterns that suggest how the environment might be maintaining or preventing a behavior. For example, a behavior analyst may notice that when a mealtime is particularly noisy, the child leaves the table and eats less than at quiet mealtimes. Or, they may find that when a parent is providing heaping spoonfuls of food, the child is more likely to gag and refuse the food. Observations about these components of the environment are important in making decisions about the intervention.

There are functional relationships between behaviors and the variables that maintain them (4). It is important to know *why* the behavior is occurring before intervening to change it. In the field of ABA, all behavior is seen to have communicative intent. For example, a child who is swatting away the spoon might be communicating that they are not hungry or that they do not want that particular food. Challenging behavior cannot just be eliminated; an appropriate replacement behavior must always be chosen. In order to do so effectively, it is crucial to determine the function of a behavior.

Functional behavior assessments (FBA) should also be conducted before implementing any intervention in order to determine why a challenging behavior is occurring. In ABA, there are various strategies and procedures of FBA. We will present an overview of the general process in this chapter, but it is a behavior analyst's job to choose the appropriate method for each individual case. In an FBA, data is taken on antecedents (events preceding the target behavior), as well as consequences (events following the target behavior). This can be done either by taking anecdotal data through observations or by manipulating antecedents and consequences in order to determine the effect of manipulations on the behavior. The latter will be discussed briefly here but should be reserved for use only in situations where other methodology is unsuccessful. It should only be conducted by professionals trained in the process.

Anecdotal FBA is commonly used as a first step before developing the intervention. Behavior analysts commonly take what is referred to as ABC data, looking for **A**ntecedents, **B**ehaviors and **C**onsequences. The antecedent refers to environmental events and conditions that occur before a challenging behavior. The consequence refers to environmental events and conditions that occur immediately after a behavior occurs. Looking for patterns in antecedents and consequences can provide insight into what is causing the persistence of the behavior and allows the behavior analyst to write a behavioral hypothesis, or a data-based idea of the environmental conditions that are maintaining the behavior. For example, if a child consistently bats a spoon away when it is presented in front of their face and this is followed by the caregiver removing the spoon, the behavior is thought to be maintained by escape from the demand of eating. If the child spits out food and the consequence is consistent reprimands from the caregiver, the behavior is hypothesized to be maintained by attention.

The benefit of conducting FBAs is the ability to determine how the environment is maintaining interfering mealtime behaviors. This information can be used to determine an appropriate function-based intervention to reduce challenging behaviors, as well as to identify appropriate replacement behaviors to meet the needs of the child. For example, a goal of intervention could be to decrease pushing the spoon away and crying, while increasing the use of language to ask for a break or to say "no, thank you."

Child and Caregiver Interactions During Mealtime

Before discussing intervention methods used in a behavioral approach, it is important to stress that both the child and feeder behavior should be assessed and considered. Descriptive information on both the feeder and child behavior can be collected in a direct observation (5). A goal of this is to look for discrepancies between the caregiver's account of the behaviors in the interview and the actual behavior during the meal. Resolving such differences could lead to improved intervention outcomes (5). Borrero et al. described a range of parent behaviors in a descriptive analysis of multiple parent-child dyads (8). Observation can lead to the identification

of parent behavior that maintains undesirable mealtime behaviors. Examples include coaxing, threatening to take away preferred items, removing the spoon or the cup from the child's mouth, providing attention in various forms, presenting preferred foods, or terminating meals following refusal (8). Borrero et al. hypothesized that caregiver responses might depend on the severity of challenging behavior (8). For example, more severe refusal behaviors such as self-injury or aggression may lead to quicker termination of mealtimes. Identifying variables that maintain both child and parent behaviors is important for the development of interventions.

In a 2006 study by Casey and colleagues, direct observation methodology was used to identify the consequences provided by caregivers (9). This study found that caregivers were not providing much positive interaction for acceptance behaviors, while frequently allowing escape from mealtime demands for refusal behaviors. This finding allowed the authors to put an intervention into place to increase the reinforcement for desirable mealtime behaviors and remove reinforcement for undesirable mealtime behaviors, leading to overall increased consumption of food and decreased inappropriate behaviors (9).

Behavioral assessment is an effective tool for selecting and defining target behaviors. Assessment leads to the identification of target behaviors to decrease, as well as appropriate replacement behaviors. It allows for the identification of prerequisite behaviors or skills that may need to be taught. Involving caregivers can assist with the prioritization of behaviors to target based on their social significance to the family. Intervention goals can be set based on this information. This also allows for the consideration of the intensity of intervention necessary. For example, a more intensive intervention may be warranted if a child and family are severely impacted by the behavior (e.g., high family stress, inability to participate in social activities, and negative health outcomes). Finally, the initial behavioral assessment allows for hypotheses to be formed about the variables maintaining challenging behavior (6).

Grace collected data across multiple mealtimes in order to operationally define the behaviors of interest, understand the interactions between child and caregiver behaviors, and look for patterns in behavior. Components of Grace's findings are provided below.

Duration of Out of Seat Behavior

Date/ Setting	Number of times leaving the table	Length of mealtime	Duration at the table	Duration away from the table	Percent of time away from the table
February 3 Home	7	34 minutes	10 minutes	24 minutes	71%
February 3 Daycare	5	18 minutes	15 minutes	3 minutes	16%
February 4 Home	6	26 minutes	11 minutes	15 minutes	58%

Number of Food Refusals

Date	Number of times caregiver offered foods	Number of acceptances	Number of refusals	Percent of refusal
February 3 Home	23	3	20	87%
February 3 Daycare	32	5	27	84%
February 4 Home	28	2	26	93%

Antecedent Behavior Consequence (ABC) Data

Date/time/ setting	Antecedent	Behavior	Consequence
2/3 9:07am Home breakfast	Dad says “take a bite of your applesauce” and puts the bowl in front of Ravi	Yells “no!” and pushes the applesauce	Dad moves the applesauce bowl away
2/3 9:08 am Home breakfast	Dad repeats “take a bite of your applesauce”	Yells “no!” turns head, and cries	Dad says “ok, ok, just drink your milk”
2/3 9:10 am Home breakfast	Mom asks “Ravi, can you please just try a bite of your egg?	Screams and runs away from table	No response
2/3 9:11 am Home breakfast	Mom brings bite of egg to living room and says “just one bite”	Screams and hits plate	Mom says “Ok, just come back and drink your milk”
2/3 11:15am Daycare snack	Daycare staff puts a carrot on Ravi’s plate	Ravi throws the carrot on the ground	None (nobody picks it up or offers him another carrot)

Grace also collected data on the types of food Ravi ate without refusals, the differences in his behavior when different caregivers presented food, the amount of food he consumed, and other environmental variables (e.g., seating arrangements, noise, distractions, etc.).

Choosing Meaningful Goals

After a thorough assessment process has been conducted, meaningful goals for intervention can be chosen. Social significance is of the utmost importance to behavior analysts when deciding on intervention goals after the assessment process. In order to develop and prioritize goals for behavior change, behavior analysts must ask themselves and stakeholders (e.g., the child, parents, caregivers, teachers, etc.) a series of questions to ensure that the goals are meaningful. Goals to prioritize are those which lead to improvements in quality of life (11). The behavior analyst should include the child (when possible) and family members in the development and prioritization of goals. Multiple considerations should be made before selecting goals for intervention.

First, the behavior analyst must ask if the goal will actually address the child's needs. It is important to distinguish between whether the child themselves will benefit or just those around them. For example, a case can be made for a child benefitting from learning to self-feed. This will benefit the child themself because they will not be dependent on others to feed them. In thinking about whether the child will benefit from the goal, it is important to ask if the current behavior poses any danger or harm to the child or others. Behaviors that will impact health or safety should be prioritized for intervention. For example, decreasing the use of a bottle might need to be prioritized for dental health. Or, increasing the variety of foods consumed might need to be prioritized for nutritional health.

Another question a behavior analyst will ask themself is if the goal is a prerequisite to other important behaviors or if it will increase the child's access to environments where other skills can be learned. For example, while learning how to sit at the table for the duration of a meal might not have clear health benefits, it is a prerequisite for being able to participate in school mealtimes in the cafeteria. School mealtimes are important for learning other skills, such as social interactions and communication. Further, a behavior analyst should be thinking about whether the goal will lead to reinforcement in the client's natural environment after the intervention ends and whether the change from intervention will be long lasting. Goals that lead to long lasting change should be prioritized over those that are temporary.

Finally, in the case of behavior reduction goals, the behavior analyst will ask themself what adaptive behavior will replace any behavior that is being reduced. As discussed previously, it is not sufficient to just reduce or eliminate a behavior without working on an adaptive replacement behavior. For example, if the behavior analyst decides to target the behavior of leaving the table during mealtime, they must provide an alternative behavior for the child, such as asking for a break or communicating their preferences around food.

In addition to the considerations above, the behavior analyst must explore cultural considerations when choosing a behavior to target. To be culturally responsive, a behavior analyst must be aware of all of the different components that make up a child's culture (e.g., family structure and values,

race, religion, ethnicity, etc.). A conversation with the family about their priorities and values is crucial when choosing meaningful goals. For example, a family might not prioritize sitting at the table for mealtime and such a goal might be inappropriate for an individual family. Likewise, a goal of decreasing screen time during meals would be inappropriate for a family who watches television together while eating.

After analyzing the assessment data and talking to Ravi's caregivers and pediatrician, Grace and Ravi's parents and caregivers came up with the following mealtime goals to be met within six months. Grace's rationale for prioritizing these goals is also provided.

1. *Ravi will stay seated at the table for one consecutive 15 minute period per mealtime. [Prioritized for Ravi to participate in home and school routines and have an opportunity to work on social and language development]*

2. *Ravi will interact with one food in a new way (e.g., touch, lick, bite, eat) three times per week. [Prioritized as a prerequisite skill for Ravi to add new foods to his repertoire]*

3. *Ravi will eat one serving size of a fruit or vegetable each day. [Prioritized for his nutritional needs]*

Grace continued to assess progress and change goals as necessary throughout the intervention process.

Behavior Change Intervention Procedures

As with assessment, intervention decisions should be individualized for the unique needs of each child and family. The following is an overview of behavioral interventions that are commonly applied to mealtime, but ultimately, it is the behavior analyst's role to choose interventions based on data gathered during the assessment and measure progress to determine if the intervention is effective.

Antecedent-based Strategies

Before intervening on the behavior itself, antecedent interventions should be considered. In an antecedent-based intervention, something in the environment is manipulated to decrease the likelihood of challenging mealtime behaviors. Antecedent-based interventions are designed to be preventive and are ideally in place *before* the behavior occurs. Sometimes, antecedent-based interventions are put in place to prevent a challenging behavior from escalating. Antecedent interventions should be directly connected to the results of the assessment. A primary goal is to make sure the mealtime environment is positive rather than aversive. The behavior analyst will think about who is present, what sensory aspects (e.g., sights, sounds, smells) are in place, and when mealtime is happening, making sure the child is motivated to participate in the mealtime routines.

Environmental Arrangement and Food Presentation

Behavior analysts might consider environmental factors, such as seating arrangements and sensory aspects, before even thinking about the food that is presented. The goal is to make the mealtime environment motivating and inviting to set a child up for success. One way to do this is to pair mealtime with highly preferred items and activities. For example, every time 3 year old Carson ran away from mealtime, he tried to play with his favorite train toys. The behavior analyst decided that she would set up mealtime with the trains already at the table, decreasing Carson's motivation to leave and increasing the likelihood of him staying at the table. Eventually, the behavior analyst might include an intervention in which the trains are gradually removed from the table. Other ways of pairing include using fun utensils and plates or making the food itself fun and appealing to the child.

After considering factors such as seating arrangement and sensory aspects of the environment, the method in which food is presented should be considered. Texture, taste, composition, and even brands and packaging are important to consider in the presentation of food (5). While some children may prefer crunchy foods, others may only be able to tolerate purees. Initially, presenting foods in ways that are preferable to the child can decrease the probability of food refusal behaviors.

Behavior analysts may also consider the quantity of food presented. Some children consistently reject food when the quantity on the plate or spoon is large , but are more likely to accept foods when a spoon is just dipped in a puree or one small bite at a time is presented (5). Pacing is another component of food presentation. Caregivers often present food at a rapid rate. When the pace of bite presentation is slowed down, some children's acceptance may increase (5). Utensils used can make a difference. For example, the use of a Nuk brush has been effective in reducing expulsion when used in place of a regular spoon (12,13). Using the Nuk brush allows therapists or caregivers to distribute food around in the mouth, taking out the extra step for the child.

Stimulus Fading

Stimulus fading is when the presentation of food is changed over time (14,15). Stimulus fading procedures are a strategy to blend preferred and non-preferred textures and flavors. In stimulus fading interventions, textures and flavors are gradually changed over time (16). Luiselli and Luiselli discussed the use of a fading procedure in which thickening agents and food particles are slowly introduced and gradually increased in a puree or smooth food, such as yogurt (5). Composition is gradually increased until the child is accepting coarser textures. Quantity can also be faded by first presenting a taste on the spoon and gradually increasing the quantity of food on the spoon (5). Bachmeyer, Gulotta, and Piazza faded from juice to applesauce by gradually adding small amounts of applesauce onto a spoon with juice (17). In another study, the distance at which a therapist held a spoon from a child's lips was gradually faded closer to the child (18). Below you will see three scenarios in which stimulus fading was implemented.

Scenario 1: Increasing the amount of milk a child consumes.
A child likes to drink chocolate milk but does not drink plain whole milk. The amount of chocolate milk being added to the cup of plain milk over time can be gradually decreased. For instance, the behavior analyst would start with 100% chocolate milk. After the child has consumed this concentration of chocolate milk, the ratio between chocolate milk and plain whole milk might decrease, so it's 75% chocolate. The chocolate milk will continue to be gradually decreased until the child is drinking a cup of 100% plain whole milk.

Scenario 2: Increasing texture to eating apple slices
In this scenario, the goal is for the child to eat apple slices. The behavior analyst might start with the applesauce that the child is already eating and add shreds of apple to the applesauce. After the child eats bites of the applesauce with small pieces of apple, the size of the apple inclusions can be gradually increased. This process will gradually continue until the child is able to take bites and chew apple slices.

Scenario 3: Combining Preferred and Nonpreferred Foods
In this final example, the goal is for the child to eat a particular nonpreferred food, such as peas. The behavior analyst would determine what preferred food could be combined with the peas. In this example, we will say that white bread is a favorite food. A tiny piece of pea might be rolled up in the bread as a starting point. Once the child is able to successfully eat that combination, the amount of peas can be gradually increased or the amount of bread can be gradually decreased until the child is comfortable with eating peas on their own.

It is important to note that in stimulus fading, the child should be told the plan rather than the foods being combined as a surprise. A goal should always be to maintain the trust of the child. If a child is tricked by a pea being inserted into their favorite food, there is a risk that they will no longer consume that food due to the aversive experience. Stimulus fading should be done gradually enough that the child feels successful and that the mealtime experience is still positive and motivating.

Providing Choices

Providing choices can be a helpful way to increase motivation to participate in the mealtime routine. When thinking about providing choices, it is important to keep a child's strengths, interests, and triggers in mind. For example, if a child is refusing fruits and vegetables, they could be given a choice of *how* to try the vegetables. Asking a child, "do you want to touch your carrot or take a little bite today?" is less likely to lead to refusal behavior than saying, "can you try your carrot?" Children can also be given choices about what utensils to use, where to sit, and how much food to put on their plate. Choices are a helpful antecedent strategy to give children some control of the mealtime experience and reduce the pressure of eating.

Positive Reinforcement

Once the environment has been considered and arranged to promote success, other strategies can be implemented. One common strategy used in ABA is positive reinforcement. While some children may be intrinsically motivated to eat, not all children experience this motivation. In this case, positive reinforcement strategies are especially helpful. In positive reinforcement, a stimulus is added into the environment following a goal behavior to increase the likelihood that the goal behavior will happen again. For example, Carson uses his fork correctly to feed himself. His mom says, "Wow, Carson! That was so great!" Carson then increases his use of a fork for self-feeding. This means that Carson's fork using behavior was reinforced. In another example, the goal for Hannah is to stay at the table longer. When she stays at the table for five minutes, Hannah gets to watch her favorite show. In the future, this increases Hannah's time at the table. It is important to note that it is the *behavior* that is reinforced, not the individual. Further, behavior analysts need to think carefully about the use of reinforcement in order to not inadvertently reinforce food consumption. The goal is for children to become intrinsically motivated to eat and stop eating when they feel satiated. Reinforcement should be used for behaviors such as self-feeding, staying at the table, participating in the mealtime routine, and interacting with foods in new ways, rather than reinforcing the eating of a certain quantity of food.

Shaping

Shaping is a strategy that uses positive reinforcement and breaks down goals into their smaller components (6). First, the behavior analyst works with the family to determine the end goal. They then break it down into smaller, achievable steps. These smaller steps are reinforced and gradually changed towards the final goal. For example, if the goal for a child is to sit at the table longer, reinforcement might first be provided after just 30 seconds. After reaching that goal successfully, reinforcement might be delivered after a minute, then three minutes, and so on until the final goal is reached. In an example where the goal is for a child to try new foods when offered, the small achievable goals might include different interactions with food. First, the child's tolerance of the food on the plate might be reinforced. Then, the child might be asked to touch the food, smell the food, and work up to taking a bite of the food. Shaping allows the child to achieve success and gradually build towards the final goal.

Replacing Challenging Behaviors

Behavior analysts see all behavior as communication. The data taken during the assessment phase helps a behavior analyst understand what the function of the behavior is, or what the child is trying to communicate. While one goal of a behavior analyst is to decrease challenging behaviors, it is not possible to eliminate such behaviors without understanding why they occur and providing the child with an appropriate replacement behavior. If a child learns to run away from the table every time a nonpreferred food is offered, a behavior analyst might teach them the words or a sign to ask for a break or to ask to remove the food from their plate. In

order to decrease communicative behaviors, the behavior analyst must provide the child with communication that serves the same purpose.

In developing an intervention plan for Ravi and his family, Grace put the following strategies in place.

Before mealtime, Ravi received a 5 minute warning. A caregiver would say "Ravi, in 5 minutes we're going to pick some toys to bring to the table." This increased Ravi's motivation to transition to the table. Ravi was given choices about which utensils to use, where to sit, and what he wanted to put on his plate. Ravi came to the table readily, excited to make his choices.

At each meal, a new food item was presented to Ravi. A shaping procedure was used to encourage new interactions with the food. Once Ravi was comfortable with the food on his plate, he received praise and play time for new interactions, such as touching, smelling, and licking the food. Eventually, Ravi became comfortable taking small bites of new foods when they were presented. Ravi was given a way to say "no, thank you" when he wasn't ready to bite a new food and was taught to use words to communicate his preferences about food.

A stimulus fading procedure was also used to work on one specific target at a time. Grace started with fruit as a goal because it was something that Ravi sometimes liked. She combined fruit with Ravi's favorite vanilla yogurt by gradually increasing the amount she added to his yogurt.

Ongoing Assessment and Collaboration

For a behavioral approach to mealtime to be successful, ongoing assessment and collaboration are crucial. Ongoing assessment consists of continuing to take data in order to measure progress towards the agreed upon goals. Continuous data collection and progress monitoring allow behavior analysts to understand whether goals are being reached and to make quick adjustments to intervention strategies when progress is not being made.

Collaboration should be present among service providers and with the family. Even the most high quality, evidence-based interventions can fail when they are not sustainable or culturally appropriate for the family. A behavior analyst should be constantly collaborating with the family to make sure intervention strategies feel feasible and align with the family's values and beliefs. Changes should be made when the family reports that the intervention approach is not a good fit.

The mealtime experience should be looked at holistically. The treatment of mealtime and feeding challenges is often discussed as an interdisciplinary challenge. Many disciplines other than ABA are involved in the treatment of such challenges, including speech-language pathology, occupational therapy, nutrition, psychology, medical specialists, primary care, and education. A

strictly behavioral approach would miss important factors, such as sensory-based interventions, nutritional needs, comorbid medical needs, oral-motor skills, etc. A behavior analyst should be aware of the skills in their scope of competence and know when to consult with other providers. When used in collaboration with family members and other service providers, a behavioral approach to mealtime has promising results.

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Chapter 10

ENTERAL FEEDING (TUBE FEEDING)

Annette Pederson, MS, RD, CD

Enteral tube feeding is used for infants, children, and youth who have functioning gastrointestinal tracts but are unable to meet their nutrition needs orally. Conditions that may require enteral feeding are numerous:

- Gastrointestinal disorders, such as disorders of absorption, digestion, utilization, secretion, and storage of nutrients; as well as anatomic disruptions, like tracheoesophageal fistula
- Inability to meet nutrition needs orally or safely by mouth, including neuromuscular disorders, such as muscular dystrophy, spinal cord defects, and cerebral palsy; or damage to the central nervous system that causes oral- motor problems
- Increased energy/nutrition needs due to cardiopulmonary disorders and other conditions, such as cystic fibrosis, burns, cancer, prematurity, chronic lung disease, catch up growth, and malnutrition.-

Enteral feeding plays a role in both short-term rehabilitation and long-term nutrition management. The extent of its use ranges from supportive therapy, in which the tube delivers a portion of the needed nutrients, to primary therapy, in which the tube delivers all the necessary nutrients. Most children and youth who receive tube feedings can continue to receive oral feedings to fulfill the pleasurable and social aspects of eating. All infants and young children require oral-motor stimulation for developmental reasons.

Tube feeding benefits the child by improving growth and nutritional status and frequently, also improves the primary condition. By ensuring that the child's nutrient needs are being met, tube feeding can free the family from anxiety and therefore, improve quality of life. Additional benefits include improved hydration, improved bowel function, and consistent medication dosage. Tube feeding is an important therapy for the child who cannot orally feed safely and needs to be fed by enteral tube to protect their airway and prevent or decrease the risk of aspiration. Tube feeding is a safer, less expensive alternative to oral feeding than is total parental nutrition (Chapter 11) (1).

There are disadvantages to enteral feeding. If a child has gastroesophageal
reflux, aggressive enteral feeding may increase their risk of aspiration or vomiting.
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Other possible physical disadvantages are diarrhea, skin breakdown, or stoma site granulation/ infection. Mechanical disadvantages include a dislodged or occluded feeding tube (1-6).

Children and youth who are either malnourished or at high risk for becoming malnourished can

benefit from tube feeding. When one or more of the following factors are identified, tube feeding should be considered after other aggressive oral interventions have been tried (1,6):

- Inability to consume at least 80% of energy needs by mouth
- Total feeding time more than four hours per day
- Inadequate growth or weight gain for more than one month (under age 2 years)
- Weight loss or no weight gain for a period of three months (over age 2 years)
- Weight for length or Body Mass Index (BMI) less than 5th percentile for age/sex
- Triceps skinfold less than 5th percentile for age

An interdisciplinary team should decide whether or not to begin tube feeding. At a minimum, the team should include the child's caregivers, the primary physician, the surgeon, and the registered dietitian nutritionist (RDN). If the child has oral-motor feeding problems, the team should also include an occupational or speech therapist. Before tube feeding is started, the child needs a medical work-up for the following purposes:

- To rule out contraindications for enteral feeding (e.g., malabsorptive disease)
- To diagnose possible gastrointestinal problems (e.g., gastroesophageal reflux, risk of aspiration)
- To determine the optimal delivery site for the feeding (e.g., stomach, duodenum, or jejunum)
- To determine an appropriate program for oral-motor stimulation

The feeding tube may be placed through the mouth or nose, such as for gavage or nasogastric (NG) feedings. A gastrostomy is placed surgically or by percutaneous endoscopic gastrostomy (PEG). The choice of placement depends on many factors (1-6):

- Expected duration of the need for tube feeding (generally, NG feeding tubes are used for short periods of time only)
- Local resources for dealing with possible complications
- Family's ability to learn the feeding technique required by the particular placement
- Preference of the caregiver(s)

Oral-motor problems may improve with development, time, and treatment. All enteral feeding techniques are reversible. Discontinuation of enteral feedings requires the same careful planning and often the same detailed work-up that go into the decision to start it.

The remainder of this section is presented in two parts: guidelines for determining when enteral feeding is necessary (Table 10-1) and guidelines for evaluating the patient who is being tube-fed (Table 10-2). The details of the enteral feeding process, including possible complications, are discussed in Appendix B.

Table 10-1 Guidelines for Determining When to Use an Enteral Tube Feeding

Assessment	Intervention	Evaluation/Outcome
Anthropometric*		
Measure and plot on appropriate growth chart: • Height or length for age • Weight for age • Weight for length or BMI for age • Head circumference (under 2 yr) **Measure:** • Triceps skinfold • Mid-upper arm circumference • Subscapular skinfold **Calculate:** • Arm muscle area • Arm fat area Obtain and plot all previous anthropometrics that are available Compare all current measurements to reference data and previous measurements	Consider tube feeding if either of the following: • Inadequate rate of growth or weight gain (for 1 month, under age 2 years; for 3 months over age 2 years), OR decreased rate of weight gain such that weight percentile has dropped continuously over past 6-12 months. • Skinfold thickness and arm fat area OR indicators of muscle mass have decreased or are below 5th percentile	Caregiver(s) and interdisciplinary team decide either to tube feed, or to continue oral feeding alone with reevaluation at later specified date Consistent growth pattern is established
Clinical/Medical		
Obtain the following: • Medical history • Review of body systems • Physical exam • Supportive laboratory work and/or X-ray (individual indications)	Consider tube feeding if any of the following: • Aspiration pneumonia (g-tube) • Anatomic abnormality in airway, upper intestinal tract, cranium, or face • Medical conditions characterized by hypermetabolic state (e.g., cardiopulmonary diseases) • Neurologic abnormality that prevents efficient oral feeding; feeding video-fluoroscopy swallowing study (VFSS) may document severity	Tube-feeding is initiated, if appropriate, without development of complications
Dietary†		
Assess dietary intake by diet history and food record Assess adequacy of energy intake based on growth records Estimate energy needs Estimate fluid needs and assess adequacy of fluid intake	Consider tube feeding if either of the following: • Oral feeding providing less than 80% of required energy • Oral feeding not meeting fluid needs	Intake of fluid, energy, protein, and other nutrients is adequate to support growth
Feeding		
Estimate number of hours per day spent feeding child. Assess oral-motor skills to determine ability to take solids and liquids Assess ability to swallow to determine risk of aspiration (e.g., VFSS)	• Caregivers spending more than 4 hr/day feeding (less time, if few caregivers) • Oral-motor skills preventing adequate oral intake of foods in a reasonable length of time • Risk of aspiration when eating or drinking	Increased time is available for parent- child interaction, without the pressure of oral feeding Oral foods offered in addition to tube feeding as appropriate for the child's swallowing ability

*For reference data and guidelines for taking accurate measurements, see Chapter 2.
†For more information, see Chapter 1 Nutrition Screening and Assessment.

Table 10-2 Guidelines for Evaluating the Patient on an Enteral Tube Feeding

Assessment	Intervention	Evaluation/Outcome
Once the decision is made to tube feed, have a gastrointestinal work-up done to document intestinal motility, anatomic integrity, and presence/ absence of gastroesophageal reflux	Determine most appropriate feeding route: • Site of formula delivery (i.e., gastric, duodenal, or jejunal) • Tube placement (i.e., nasal or surgical)	Caregiver(s) and all involved medical professionals contribute to decisions regarding feeding route
Have a medical/surgical assessment done	Determine requirements for the following: • Fluid • Energy • Protein • Vitamins • Minerals • Electrolytes	Patient is receiving a nutritionally adequate feeding
Before starting tube feeding, do a complete nutrition assessment:* • Anthropometric • Biochemical • Physical • Dietary	Determine most appropriate type of formula and supplements. Determine most appropriate method of formula delivery (i.e., bolus, continuous drip, or combination)	
	Instruct caregiver(s) about the following:† • Obtaining the formula and supplements • Preparing the formula • Giving feedings and using the pump • Daily skin/stoma care • When to call physician	Caregiver(s) obtain appropriate formula and supplements Caregiver(s) demonstrate appropriate techniques for feeding and stoma/skin care Caregiver(s) know when to call MD
Once tube feeding has begun, monitor closely: • Tolerance of tube feeding; maintain frequent contact with family by phone or clinic visits; check for vomiting, diarrhea, constipation, and other adverse reactions • Growth and indicators of fat and muscle stores, every 1 - 2 months until weight gain has been stable for 2 months	As necessary make changes in: • Type of formula • Amount of formula • Method of delivery • Additional supplements • Once child achieves appropriate weight for height (or length), evaluate need for reducing energy intake to compensate for low energy needs due to immobility or paralysis	Child tolerates feeding regimen and formula well (no gastrointestinal disturbances or other signs of formula intolerance) Weight gain is stable and adequate
Once weight gain has been stable for 2 months, re-evaluate every 6 months (more often in periods of rapid growth, such as infancy and adolescence): • Growth and indicators of fat and muscle stores • Nutrient adequacy of formula • Method of formula delivery • Tolerance of formula	Make changes in formula and delivery method as indicated by nutritional status.	Weight is appropriate for height (or length) Formula meets requirements for energy, protein, vitamins, minerals, and electrolytes Fluid intake is adequate

*For more information, see Chapter 1 Nutrition Screening and Assessment.
†See Appendix B Technical Aspects of Enteral Feeding.

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Chapter 11

COMMUNITY MONITORING OF THE PATIENT ON HOME PARENTERAL NUTRITION

Kathryn L. Hunt, RDN, CD

Parenteral nutrition (PN) is needed when the patient's gastrointestinal tract will not sustain life or when a child is unable to tolerate enteral feedings for a significant period of time. PN may be administered for weeks or months, as may be required for children with nutritional depletion from certain diseases, or for years, as may be required for children with severe short bowel syndrome (1). Although PN serves as a life sustaining treatment in situations where impaired gastrointestinal function precludes adequate enteral nutrition intake (EN), the use of enteral nutrition is the preferred route of nutrition delivery and should be used along with PN when feasible (8). Indications for home parenteral nutrition for pediatric patients include:

- Short bowel syndrome
- Intestinal motility disorders (e.g., pseudo-obstruction)
- Inflammatory bowel disease (e.g., Crohn's disease, ulcerative colitis)
- Hypermetabolic states (e.g., severe burns and trauma)
- Acute and chronic pancreatitis
- Special circumstances (e.g., hepatic failure, cancer, congenital villous atrophy)
- Unexplained intestinal malabsorption syndromes

The goal of the health care team in managing the pediatric patient on PN is to provide a solution of carbohydrate, protein, and fat that will achieve a positive nitrogen balance for growth, increase the patient's weight, and improve clinical outcome. The solution should also contain appropriate amounts of electrolytes, vitamins, minerals, and trace elements to maintain balance within the patient's body and prevent complications of nutrient deficiencies or toxicities (2). Despite its life-saving capacities, PN has risks and potential consequences which must be considered, including metabolic disturbances, liver damage from overfeeding, biliary sludge from absence of enteral nutrition, metabolic bone disease, and catheter-related complications, such as thromboses and infections (3,8,9). To minimize these risks and to prevent other complications, the patient receiving PN should be transitioned to enteral feeding at the earliest opportunity (4). This requires close monitoring and regular re-evaluation by the health care team.

In general, PN regimens (including solution composition) are established in the hospital, where the

patient's metabolic response and tolerance can be monitored closely (5). Pediatric candidates for home parenteral nutrition (HPN) must be clinically stable, and parents/caregivers must be trained and able to independently and safely perform the medically complex procedures to deliver PN at home (8). After discharge, it is important to continue to monitor response to PN and regularly re-assess nutrient needs. Guidelines for the technical aspects of PN, including complications of PN, are provided in Appendix C.

The remainder of this section presents the basic guidelines for monitoring a child on home PN. Regular assessment and monitoring by a team of health care professionals, including physician, pharmacist, and registered dietitian nutritionist (RDN) with PN experience, is essential. Coordination of care between the ordering physician and home care company is vital.

Table 11-1: Community Monitoring of the Pediatric Patient on Home Parenteral Nutrition

Assessment	Intervention	Evaluation/Outcome
Anthropometric[1] after hospital discharge		
Measure and plot on appropriate growth chart • Weight for age (weekly for infants, 2 times per month for older children) • Length for age (0-3 years) (Monthly in infants and young children) • Height for age (2 years and older) (every 3 months, depending on clinical status) • Weight for length or BMI • OFC: (monthly in infants, every 3 months in children up to 3 yrs)	• If weight loss or no weight gain, increase energy provided by PN solution. • Weight for length, weight for height, or sex less than 10th %ile • BMI for age or sex less than 5th %ile (8) • If rate of weight gain exceeds recommended guideline on 2 consecutive visits, decrease energy provided by PN solution (6).	Steady, stable weight gain to maintain normal growth curve. Consistent linear growth. Weight gain to improve weight for length, weight for height and BMI within normal parameters. Consistent increase in OFC along usual growth channel.
Measure (2 years and older) every 1-3 months, depending on clinical status: • Triceps skinfold • Mid-upper arm circumference • Subscapular skinfold Calculate: • Arm muscle area • Arm fat area	Use information in assessing child's energy and protein needs	Restore muscle and fat reserves to normal or >10th percentile
Biochemical		
Frequency of monitoring biochemical indicators depends on the child's clinical condition and the protocol established by the institution. Once the medical condition is stable, labs are generally done every 1-3 months or when PN solution is adjusted: • Electrolytes • Blood glucose • Ionized Calcium, phosphorus, magnesium • Folate • Creatinine, BUN • CBC, platelets • Prealbumin, albumin • Triglycerides • Prothrombin time • Zinc, copper, selenium, vitamin B12 • Ammonia • Liver function tests: ALT, AST, GGT, and Bilirubin (unconjugated/ conjugated) • Alkaline phosphatase • Fat-soluble vitamins (A, E, D (25-OH))	Work with PN team to monitor biochemical indicators and assess need to adjust PN solution.	Biochemical indicators in the normal range

continued...

Assessment (continued)	Intervention	Evaluation/Outcome
Clinical		
Observe child for signs of PN-related complications: • Infections (e.g., catheter or line sepsis): indicated by fever, redness at catheter site, elevated triglycerides or glucose levels, lethargy • Mechanical (e.g., catheter occlusion): indicated by clot or thrombus, failure to maintain line patency, formation of fibrin sheath outside catheter, fat deposition or mineral (calcium and phosphorus) precipitates • Metabolic abnormalities (e.g., electrolyte imbalances, glucose instability, elevated triglycerides, elevated liver function tests) indicated by abnormal lab values	Immediately alert physician and PN team of signs of complications	Complications are identified and treated
Dietary/Feeding		
Assess feasibility of enteral (tube-feeding or oral) trial. Evaluate: • Developmental readiness • Medical readiness • Readiness of family • Level of oral stimulation	Make referral to occupational therapist in advance of starting enteral feeding, for oral stimulation, prevention of future feeding aversions Gradually begin oral or tube feeding when feasible	Transition to enteral (tube or oral) feedings is begun (or postponed)
Typically, PN is weaned in a balanced way across the three substrates (dextrose, protein and lipids) as enteral or oral nutrition advances and is tolerated. During transition, monitor weight closely.	As enteral intake approaches 50% of requirements for energy, protein, and micronutrients, and is absorbed by GI tract, begin decreasing energy provided by PN solution (8). Consider: • Reduce rate of PN infusion by 1 mL for every 1 mL tube feeding rate increase • Eliminate lipid infusion when 50-60% energy needs are met enterally and weight is stable • Reduce number of hours of PN infusion • Discontinue PN when 75-80% energy needs met orally or enterally and there is adequate nutrient absorption (8)	Fluid, energy, protein, and micronutrients provided by parenteral and enteral nutrition meets child's estimated needs for growth and weight gain

[1] For reference data and guidelines for taking accurate measurements, see Chapter 2.

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Chapter 12

ACCOMMODATING CHILDREN WITH SPECIAL DIETARY NEEDS IN SCHOOL NUTRITION PROGRAMS

Betty Marcelynas, MA, RD and Bette Brandis, RD Updated by Donna Parsons, MS, RD
Updated 2023 by Ben Atkinson, MS, RD, CD

It has been estimated that 18% of all children and youth have special health care needs (1). Case studies conducted in eight school districts have shown that most school food service managers serve at least one student with special nutrition needs in their school on a regular basis (2).

Some students attend special education classes due to developmental disabilities, while others with arthritis, pulmonary disease, food intolerances, diabetes, and inborn errors of metabolism (e.g. PKU, galactosemia) may attend regular classes. The medical conditions reported most frequently by schools include food allergies, diabetes, and a variety of disorders that require modifications to the texture of food (2).

Legislation

Several federal laws have been passed with the intent of ensuring that all enrolled students, regardless of disability, have access to meals served at school and, that those students eligible for free or reduced-price meals receive them. Under Section 504 of the *Rehabilitation Act of 1973* and the *Americans with Disabilities Act* (ADA), a "person with a disability" is defined as anyone who has a physical or mental impairment that substantially limits one or more major life activities, has a record of such an impairment, or is regarded as having such an impairment (3).

Major life activities covered by this definition include caring for one's self, eating, performing manual tasks, walking, seeing, hearing, speaking, breathing, learning, and working (3).

One effect of these laws and the *Individuals with Disabilities Education Act* (IDEA, 1990) has been an increase in the number of children with disabilities who are being educated in regular school classrooms. Often, the disability prevents the child from eating meals prepared for the general school population. Students with special nutritional needs usually have the same or greater nutritional needs as students without physical disabilities; however, they may have a difficult time meetings those needs (4).

During the SARS CoV-2 (Covid-19 disease) outbreak, the USDA allowed school districts to provide free breakfast and lunch for all students, regardless of household income level. In 2021 and 2022, some state legislatures passed laws to fund free meals for all students, removing previous barriers to families and students. In Washington State, the legislature passed house bill 1878 to increase the number of schools able to provide free breakfast and lunch by increasing funding to the Community Eligibility Provision (5).

School Food Service Requirements

Students who may need modified or special meals can be classified in two major categories: 1) those who are unable to eat regular school meals because of a disability and 2) those who have a chronic medical condition but are not determined to have a disability. A sample order form that a physician or recognized medical authority may use for students with disabilities or a chronic medical condition to modify a diet is included (Appendix D).

Students with Disabilities

The US Department of Agriculture's (USDA) nondiscrimination regulation (6), as well as the regulations governing the National School Lunch Program (7) and School Breakfast Program (8), make it clear that substitutions to the regular meal must be made for students who are unable to eat school meals because of their disabilities when that need is certified by a statement or order signed by a recognized medical authority.

The order must include (3):

- The student's disability and an explanation of why the disability restricts diet
- The major life activity affected by the disability
- The food or foods to be omitted from the student's diet and the food or choice of foods to be substituted

Students with Chronic Medical Conditions

For a student without a disability, but with a chronic medical condition that requires a special diet, an order signed by a recognized medical authority must be provided. (In Washington State a recognized medical authority is defined as a Medical Doctor (MD), Doctor of Osteopathy (DO), licensed Physician's Assistant (PA) with prescriptive authority, an Advanced Registered Nurse Practitioner (ARNP), or a licensed Naturopathic Physician).

This order must include:

- Identification of the medical or other special need which restricts the child's diet
- The food or foods to be omitted from the child's diet and the food or choice of foods that may be substituted (e.g. texture changes and foods substitutions) (7)

Other items that may be included in orders for children with disabilities or chronic medical conditions are:

- Whether the allergy/medical condition is temporary or permanent. (A permanent note will relieve the family from updating this information every year.)
- The location for maintaining this documentation to ensure that it accompanies the student should she transfer to another school or district

Students with Other Special Dietary Needs

Schools may make food substitutions, at their discretion, for individual students who do not have a disability, but who are medically certified as having a special medical or dietary need. Such determinations are made only on a case-by-case basis and must be supported by a statement or order that specifies the food substitution needed and is signed by a recognized medical authority.

This provision covers those children who have food intolerances or allergies, but do not have life-threatening reactions (anaphylactic reactions) when exposed to the foods to which they are allergic. Generally, children with food allergies or intolerances do not have a disability as defined under USDA's regulations and school food authorities may, but are not required to, make substitutions for them.

However, when in the physician's assessment, food allergies may result in severe, life-threatening (anaphylactic) reactions, the student's condition would meet the definition of disability and the substitutions ordered by the physician must be made (6).

Schools are not required to make modifications to meals due to personal opinions of the family regarding diets.

Students with Individualized Education Plans

Many students with special needs will have an Individualized Education Plan (IEP) or an Individualized Family Service Plan (IFSP). These are plans for students receiving special education and related services to help the student benefit the most from the school program. The services described in the IEP or IFSP may include special meals, supported by a diet order. The food service director or manager is responsible for providing meals as described in the diet order, but is not responsible for revising, changing, or interpreting the diet order (7). Examples of IEP nutrition-related goals that are written in collaboration with the child's nutrition team and parents are outlined in Appendix E.

Section 504 of the *Rehabilitation Act of 1973* specifies that food service program administrators must serve special meals at no extra charge to students whose disability restricts their diet (8). There is no provision for additional federal reimbursement for the added expense.

However, these costs are legitimate program costs that can be paid for out of the food service account, which includes federal reimbursement for meals served for these students. If federal reimbursements are insufficient, alternative funding sources may also be available from Medicaid and special education to cover some of these costs. School officials should explore all possible funding sources.

Nutrition Team

The team for a student with special nutritional needs often includes the principal and teachers, the food service director and/or staff, the child's parents, and other health professionals and specialists. The team considers the needs and abilities of the individual student. The food service staff:

- applies basic guidelines for food preparation to meet those needs
- refers to resources, including parental input, on the proper techniques for preparing regular menus in a special way
- with the rest of the team, evaluates whether or not the meal plan is meeting the special needs of the student

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Chapter 13

NUTRITION INTERVENTIONS FOR CHILDREN WITH ATYPICAL GROWTH PATTERNS

Mari Mazon, MS, RDN, CD

Assessing the nutrition status of children who grow differently from the general population can be challenging. The development process of tools such as growth charts and guidelines for identifying malnutrition exclude children with diagnoses that affect growth (1). Children may grow differently due to chromosomal changes or lack of ability to ambulate or bear weight (2). Examples of conditions that result in an atypical growth pattern are listed in Table 13-1.

Obtaining a full set of anthropometrics, beyond just weight, is extremely important for conducting nutrition assessment in children with an atypical growth pattern. Children with significantly short stature may appear malnourished based on weight information alone. Attempting to have such children gain weight may result in overweight or needless pressure placed on caregivers that may disrupt the feeding relationship. Children who are non-ambulatory have low muscle mass and may appear underweight on a growth chart. Nutrition interventions to increase their weight-for-length or body mass index (BMI) may lead to increased adiposity.

Weight-for-length or BMI may not accurately describe a child's muscle or fat status particularly in children with conditions that may alter body composition or body proportions (1, 3, 4). Evaluating mid-upper arm circumference, triceps skinfold, or subscapular skinfold in addition to weight-for-length or BMI provides a fuller picture of a child's nutrition status (1, 3). Monitoring trends over time is more useful than comparing a single measurement to a reference standard developed for the general population (3). See Chapter 2.

Specialty growth charts should be used with caution. Due to the rareness of these conditions, they are often based on a small sample size and a non-representative sample of convenience (2). Children included in the development of these growth charts may have had feeding difficulties that compromised their nutrition status. Such growth charts describe the growth pattern seen in a population and not how they should grow. For example, a child within a "healthy" BMI range on a Prader-Willi syndrome growth chart may flag for overweight on a growth chart for the general population. For this reason, the Centers for Disease Control and Prevention (CDC) recommends that specialty growth charts be used along with standard growth charts such as the World Health Organization for birth to 24 months and CDC for ages 2 to 20 years (1-3). Monitoring weight-for-

length or BMI trends on a standard growth chart can be helpful for screening for overnutrition in children with low stature (5).

Table 13-1: Diagnoses Associated with Atypical Growth Patterns

Diagnosis	Growth Pattern	Specialty Growth Chart
Down syndrome	Short stature	Sex-specific[6] 0-36 months • Weight • Length/height • Weight-for-Length • Head circumference 2-20 years • Weight • Height • Head circumference
Prader-Willi syndrome	Short stature	Sex-specific Growth hormone-treated[7] 0-18 years • Weight • Length/height • Weight-for-Length/BMI • Head circumference Non-growth hormone-treated[8] 3-18 years • Weight • Height • BMI • Head circumference
Turner syndrome	Short stature	Non-growth hormone-treated[9] 2-19 years • Height
Achondroplasia	Short stature, large head, proportionately short arms and legs	Sex-specific[4] 0-20 years • Weight • Length/Height • Head circumference • BMI • Waist circumference • Head circumference • Sitting height • Sitting height:height • Arm span • Leg length • Foot length
Noonan syndrome	Short stature	Sex-specific[10] 0-36 months • Length 2-20 years • Height • Growth velocity

Cerebral palsy	Decreased fat, decreased muscle	Sex-specific[11] GMFCS* specific Tube fed/non-tube fed 2-20 years • Height • Weight • BMI
Spina bifida	Short stature, decreased muscle	Not available

*Gross motor function classification system

In addition to anthropometric information, visual assessment and nutrition-focused physical exam provide important information regarding a child's nutrition status (1). It is helpful to visualize muscle and adiposity in all four extremities and trunk. This may require young children to be in their diaper or underwear only and older children to be wearing loose, light clothes that can easily expose these areas. Visual assessment is especially useful when length or height measurements are not available to assess weight-for-length or BMI, or when assessing children with uneven fat distribution or atypical body proportions.

Children with atypical growth patterns often have different energy requirements compared to the general population. Children with low stature, low muscle mass, gross motor delays, or limited mobility may have lower energy needs. Standard energy estimation equations will overestimate their needs. Energy estimation equations for specific diagnoses exist but should only be used as a starting point, followed by close growth monitoring (3, 12). For some conditions that require very low energy needs, kcal/cm height may provide a better estimate than kcal/kg weight (3, 12, 13). Assessment of protein, micronutrient, and fluid adequacy is very important for children with very low energy needs to ensure they are meeting the recommended dietary allowance for age (12, 14).

The remainder of this chapter presents guidelines for nutrition assessment, intervention, and evaluation/outcome for children with atypical growth patterns.

Table 13-1: Nutrition interventions for children with atypical growth patterns

Assessment	Intervention	Evaluation/Outcome
Anthropometric*		
Measure and plot on standard growth chart and specialty growth chart, if applicable: • Length/height for age • Weight for age • Weight for length or BMI • Head circumference Additional measurements that may be useful: • Mid-upper arm circumference • Triceps skinfold • Subscapular skinfold • Knee height/tibial length if cannot measure height (14) Obtain and plot previous anthropometric data available. Compare all current measurements to reference data and to previous measurements. Note visual signs of nutrition status (adiposity, muscle mass, signs of wasting) Collect data from medical records, caregivers, and child (if possible): • Growth history • Growth velocity	If weight-for-length/BMI z-score or percentile is increasing rapidly or excess adiposity is determined upon visual assessment/skinfold measure, dietary or family/social interventions that result in the following is appropriate: • Weight maintenance or reduced rate of weight gain until weight-for-length/BMI z-score or percentile stabilizes (15). For a child with weight-for-length for BMI >95th percentile on WHO or CDC growth chart and trending upward, dietary or family/social interventions that result in • Weight maintenance or weight loss <1 lb/month until BMI returns to previously established channel is appropriate. Children 12-18 years of age may safely lose up to 2 lbs/week (15). For a child who is underweight based on • Weight-for-length or BMI z-score <-1.0 and trending downward (16) • Mid-upper arm circumference z-score <-1.0 and trending downward (16) • Visual assessment Dietary or family/social interventions that result in an increased rate of weight gain or weight-for-length, BMI, or mid-upper arm circumference z-score >-1.0 is appropriate.	Monitor growth monthly until weight-for-length or BMI follow a consistent pattern (15). Monitor growth monthly until no visual signs of malnutrition.
Clinical/Medical		
In absence of obvious nutrition/lifestyle contributors to growth trend, consider underlying medical reasons.	Refer to primary care provider.	Underlying medical reasons for growth pattern are ruled out or addressed.

Determine current activity level and physical capability for increasing activity. Physical activity helps increase lean body mass, decrease fat mass, and improves mental and physical health. Families of children with atypical growth patterns may need help identifying safe physical activities for their child.	If child is ambulatory and able to follow instructions, develop with caregivers and child a plan for increasing daily activity.17 Consider: • Activities the child enjoys • Involving family or friends • Local parks and recreation programs specifically for children with physical and intellectual disabilities • Mix of weight-bearing and aerobic activities (~30 minutes) • Special Olympics program for children >8 years of age (Special Olympics Young Athletes program for 2.5-7 year-olds) If child is in wheelchair or has some degree of physical disability, refer to physical or occupational therapist regarding a plan for engaging in physical activity. If child is quadriplegic or unable to move voluntarily, engaging in physical activity may not be an option. See Chapter 3 for more information about physical activity.	Caregivers, educators, and others involved in child's daily care report participation in physical activity.
Dietary†		
Obtain detailed nutrition information by interviewing caregivers and child (if possible). • Time of snacks and meals • Food preparation methods and portions • Beverages consumed • Access to food away from home (school, day care, other homes, stores) • Use of food as rewards • If tube fed, assess fluid flushes and any oral intakes Obtain a 3- to 7-day food record. Calculate average daily energy, protein, and micronutrient intakes (iron, zinc, calcium, vitamins C, D).	Calculate energy intakes and determine kcal/cm height or kcal/kg weight. Estimate energy needs based on current intakes and growth pattern. (Examples of energy needs based on diagnosis: *Prader-Willi syndrome* –8-11 kcal/cm for maintaining growth in channel; 6-8 kcal/cm for gradual weight loss (13) *Spina bifida* -- 9-11 kcal/cm for maintenance; 7 kcal/cm for gradual weight loss) (3)	Subsequent food records or interview show intakes within desirable kcal/cm height or kcal/kg weight range. Re-evaluate nutrition care plan according to changing energy needs due to growth, changes in body composition, and activity level.

continued...

Assessment (continued)	Intervention	Evaluation/Outcome
	Develop individualized nutrition care plan based on the following: • Estimated energy needs • Child's food likes, dislikes, allergies, and intolerances • Family's and child's eating patterns • Child's feeding skills and mode of feeding (oral or tube) • Family's financial resources Encourage positive feeding interactions, support for self-regulation of food intake, allowing for choices, and avoiding unnecessary rigidity or "rules" related to food (18) Help design reinforcement strategies at home and school that do not involve food Obtain periodic food records or diet history as indicated. Assess average daily energy intake.	
Family/Social		
Determine: • Family's feelings about child's growth • Previous attempts to control growth • Family's pattern of reinforcement for eating or any use of food as rewards	If family has growth expectations that may not be achievable based on child's diagnosis, communicate with all health care providers to agree on an appropriate message to be reinforced by all.	Family and health care team agree on growth expectations.

*For reference data and guidelines for taking accurate measurements, see Chapter 2.
† For more information about dietary assessment, see Chapter 1.

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Chapter 14

NUTRITION INTERVENTIONS FOR THE PREMATURE INFANT AFTER DISCHARGE

Joan Zerzan, MS

Advances in neonatal intensive care, including respiratory management and nutrition support, have contributed to a dramatic increase in the survival of infants born prematurely and of low birth weight (LBW). See Table 14-1 for definitions. These infants may experience prolonged hospitalization and medical and nutritional complications commonly associated with their immaturity. Increasing numbers of "late or moderately preterm infants" are being discharged home before their due date. However, considerable diversity exists in this population. Some infants are discharged without any major medical complications related to prematurity. Others experience continuing health problems and emerging neurodevelopmental sequelae after discharge from the hospital. Medical and developmental complications present at discharge or that emerge in the post discharge period may further impact the needs of individual infants (1-6). Management of the individual infant requires a thorough assessment of nutrition status with a focus on the Nutrition Care Process.

Table 14-1: Categories of Infants by Birth Weight and Gestational Age

Term	Definition
Prematurity	<37 weeks gestation
Late or moderately preterm infant	32-<37 weeks gestation
Very preterm	28-<32 weeks gestation
Extremely preterm	<28 weeks gestation
Low birth weight (LBW)	≤2500 grams or 5.5 lbs
Very low birth weight (VLBW)	≤1500 grams or 3.3 lbs
Extremely low birth weight (ELBW)	≤1000 grams or 2.2 lbs
Small for gestational age (SGA)	Birth weight <10th percentile
Appropriate for gestational age (AGA)	Birth weight 10th-90th percentile
Large for gestational age (LGA)	Birth weight >90th percentile

Medical and Developmental Factors

Preterm infants exhibit increased morbidity and mortality when compared to term infants (1-6). The risk for increased morbidity and mortality is inversely related to gestational age and size. Morbidity associated with the late preterm infant includes temperature instability, hypoglycemia, respiratory distress, feeding difficulties, and growth failure in the immediate post discharge period. Compared to term infants, the late preterm infant is at increased risk of negative neurodevelopmental outcomes (4-6). Complications that may affect nutritional needs and growth outcomes are listed in Table 14-2.

Table 14-2: Complications That May Affect Nutritional Needs and Growth

Physiological System Affected	Possible Complications	Reference in this volume
Respiratory	Bronchopulmonary dysplasia (BPD) Reactive airway disease (RAD)	Chapter 15
Cardiac	Congenital heart disease (CHD) Patent ductus arteriosus (PDA) Cor Pulmonale	Chapter 18
Renal	Nephrocalcinosis	
Gastrointestinal	Gastroesophageal reflux (GER) TPN-induced cholestasis Short bowel syndrome (SBS)	Chapter 8 Chapter 11 Chapter 18
Neurodevelopmental	Developmental delays Cerebral palsy Learning disabilities Vision and hearing	Chapter 8 Chapter 8
Hematological	Anemia	
Immunological	Susceptibility to repeat infections, illnesses, and rehospitalizations	
Nutrition	Alteration in growth including postnatal growth restriction and malnutrition Osteopenia Feeding difficulties Other nutrient deficiencies	Chapter 2 Chapter 5 Chapter 8

Growth Expectations and Assessment

During hospitalization, it is common practice to strive for “in utero” rates of growth (15 g/kg/day weight gain, 0.5-1.0 cm/week increase in length). After hospital discharge, it is unclear what represents optimal growth for the individual preterm infant. Factors such as growth history and postnatal growth restriction have implications for growth expectations. A number of growth charts have been developed for monitoring growth during hospitalization. These charts have been designed to reflect either intrauterine growth standards or postnatal growth patterns. The INTERGROWTH-21st growth study allows for growth monitoring to 64 weeks postmenstrual age (PMA).

The WHO growth charts developed for term infants are also used for monitoring growth of preterm infants after discharge. It is usual practice to use the WHO growth charts to follow the growth of preterm infants from 40 weeks PMA following discharge (7-12).

When using the WHO growth charts, growth should be plotted according to corrected age until approximately 2 years of age (4). Weight gain velocity tables from WHO data can be used for the preterm infant adjusting for corrected age (See tables 14-3 and 14-4. Also available at https://www.who.int/childgrowth/standards/w_velocity/en/).

See example below for calculating corrected age:

> Corrected age (CA) = Chronological age (CH) – number of weeks premature
> Example: An infant is born at 28 weeks gestation and is now 6 months past their date of birth:
> Step 1: 40 - 28 = 12 weeks or 3 months premature
> Step 2: 6 months - 3 months = 3 months CA

Table 14-3: Weight Gain Velocity for Males (grams/day)

Age in Months CA*	3rd-5th percentile	15th-25th percentile	50th percentile	75th-85th percentile	95th-97th percentile
0-3	23-33	23-36	26-39	30-39	30-46
3-6	10-20	13-20	13-20	13-23	16-23
6-9	7-10	7-10	10-13	10-13	10-16
9-12	3-10	7-10	7-10	7-10	10
12-15	3-7	3-7	7-10	7-10	7-10
15-18	3-7	3-7	7	7-10	7-10
18-21	3-7	7	7	7-10	7-10
21-24	3-7	3-7	7-10	7-10	7-10

Table 14-4: Weight Gain Velocity for Females (grams/day)

Age in Months CA*	3rd-5th percentile	15th-25th percentile	50th percentile	75th-85th percentile	95th-97th percentile
0-3	20-26	20-30	23-33	26-36	30-39
3-6	10-16	10-16	13-20	13-20	16-23
6-9	7-10	10	10	10-13	10-16
9-12	3-7	7	7-10	7-10	10
12-15	7	7	7-10	7-10	7-10
15-18	3-7	3-7	7	7-10	7-10
18-21	3-7	7	7-10	7-10	7-10
21-24	3-7	3-7	7	7-10	7-10

* CA = corrected age

(Danner E, et al. Weight velocity in infants and children. Nutr Clin Pract. 2009;24:76-69)

Postnatal growth restriction remains a serious problem for preterm infants and follow-up studies suggest that when growth parameters are plotted according to corrected age (CA), VLBW and ELBW infants may not achieve percentiles comparable to term infants of similar age; they remain smaller and lighter. Most studies demonstrate little "catch-up growth" for the VLBW infant between 1-3 years of age. Infants with chronic medical conditions may not experience "catch-up growth" until school age (7, 8). It is important to note that the term "catch-up growth" is often used in a non-traditional sense to identify infants who achieve ≥ 10th percentile on growth charts. More correctly, "catch-up growth" describes an infant who demonstrates accelerated rates of growth following a period of growth failure. The infant who continues to gain 20-30 g/day after 6 months of age, or the SGA infant who is more than twice their birth weight by 4 months of age, may be demonstrating a pattern of accelerated growth even though they remain <10th percentile in growth parameters.

VLBW and ELBW infants are also at increased risk of malnutrition (or undernutrition), previously identified as "failure to thrive" (FTT) (13-17). The term FTT in the traditional sense refers to failure to gain in weight and length at expected rates. One study identified a 21% incidence of FTT in VLBW infants in the first 36 months of life. The incidence of FTT peaked between 4-6 months of age (18-19). Preterm infants are at risk for being misidentified as having FTT when the term is applied in other ways (i.e., weight or length <5th percentile).

ELBW and infants with severe intrauterine growth restriction (IUGR) may demonstrate periods of accelerated rates of weight gain and remain ≤10th percentile in weight and length for several years. Therefore, the rate of growth and weight gain should be evaluated.

Nutrition Practices Associated with Growth Outcomes

Preterm infants are often discharged from the hospital when they weigh approximately 1800-2000 grams (4.0-4.5 lbs), are nippling all feedings, and can maintain their temperature outside an isolette. Infants who are unable to demonstrate adequate oral feeding may be discharged with supplemental tube feeding. It is usual practice to transition these infants off preterm formula and fortified breast milk. Discharge feedings include post discharge premature formula, standard infant formula, or breast/chest milk. These feedings are often augmented with additional calories. There are a variety of ways additional calories may be added: using post discharge formula, increasing concentration, or supplementing breast/chest milk feedings with additional calories.

Although some studies have demonstrated improved growth rates in infants fed a nutrient-enriched formula after hospital discharge, there is insufficient evidence to support routine use for all preterm infants. Recognition of the heterogeneity of this population supports individualization of feeding choices (20-26). Factors to consider in feeding choices for the preterm infant include individual nutrient needs, tolerance, parental choice, and cost and availability. Practices that were associated with poor growth outcomes in a group of VLBW infants include (10):

- Introduction of solids prior to 6 months CA
- Introduction of cow's milk before 12 month CA
- Use of low fat milk

Feeding Difficulties

Preterm infants who have attained an age at which oral-motor maturity supports nipple feeding may continue to have feeding issues. Factors such as immature maintenance of physiological stability, disorganized suck-swallow-breathing, decreased strength and endurance, cardiorespiratory compromise, past feeding experiences, and neurodevelopmental complications may contribute to alterations in feeding behavior and ultimately feeding success (27-30). Infants who experience unpleasant feeding experiences (choking, respiratory distress, GER) may begin to demonstrate aversive feeding behaviors. Evaluation of preterm infants with growth concerns and/or reports of feeding difficulties should include a careful history, description of feeding behaviors, and observation. In observing a feeding, attention should be given to document control, organization, coordination of suck-swallow-breathing, length of time to consume adequate volume, evidence of distress, signs of choking, and changes in respiratory status. Infants who demonstrate evidence of feeding difficulties should be referred to the appropriate disciplines for further evaluation and treatment. See Chapters 8 and 9. Interventions should focus on cue-based feeding and support development and feeding relationships.

Nutrient Needs

The nutrient needs of preterm infants after hospital discharge and throughout the first year have not been clearly established. Common practice is to view the nutrient needs of the preterm infant to be the same as the term infant when the preterm infant achieves a weight of 2.0-2.5 kg (4.5-5.0 lbs). Some follow-up studies raise questions about this practice (10-12). Infants fed a nutrient-enriched formula after discharge show improvements in growth and mineral status. Follow-up studies have also demonstrated decreased bone density in VLBW infants one year after discharge (31-33). Some preterm infants may continue to be at risk for inadequate bone mineralization after discharge. These infants may need higher mineral intake and monitoring after hospitalization. Currently there are no standardized practices to treat these infants, and a variety of strategies have been used without clear identification of an optimal approach. Often, the transition to breast/chestfeeding occurs after discharge from the hospital. These infants may continue to receive supplemental bottles of formula or breast/chest milk until the transition to total breast/chestfeeding is complete. To facilitate transition, follow-up is essential. This follow-up can be provided by a hospital or community lactation specialist.

Standard infant formulas are designed to meet the Dietary Reference Intakes (DRI) for vitamins and minerals for term infants when the infant consumes approximately 32 oz/day. Infants discharged from the hospital weighing 4.5-5.0 lbs may only consume 10-12 oz/day. This volume

may be adequate to meet fluid, energy, and protein needs. However, a multivitamin supplement is needed to meet the DRI for infants until the infant or child consumes 24-30 oz/day. Soy formulas are not recommended for preterm infants, particularly those at risk for osteopenia, secondary to decreased bioavailability of calcium and phosphorus (23).

Preterm infants often demonstrate adequate weight gain when consuming 110-130 kcal/kg/day. The VLBW and ELBW infants with ongoing medical issues may need higher energy intakes to support appropriate weight gain. Factors that alter energy needs, absorption, or utilization in infants will also impact the energy requirements of preterm infants.

Preterm infant formula and human milk fortifiers are designed to meet the increased vitamin and mineral needs of the preterm infant taking smaller volumes than the term infant consumes. Continuation of the preterm infant formula and human milk fortifiers in infants who weigh more than 2.5-3.0 kg will result in increased intakes of several vitamins, including vitamins A and D. Case reports of hypervitaminosis D suggest that these products should be discontinued when the infant is exceeding the recommended intakes for fat-soluble vitamins.

Preterm infants are at risk for iron deficiency anemia. Preterm infants require 2-4 mg iron/kg/day by 2 months of age. This may be provided as an iron supplement or with the appropriate volume of iron-fortified formula. In general, this iron should be continued until 12 months of age (CA) (23).

The remainder of this chapter presents guidelines for nutrition assessment, intervention, and evaluation/outcome after discharge from the hospital for children who are born prematurely. Additional references and resources are included at the end of this chapter.

Assessment	Intervention	Evaluation/Outcome
Anthropometric		
Measure[1]: • Length for age • Weight for age • Weight for length • Head circumference Plot on appropriate growth chart: • Premature growth curves • WHO charts using corrected age (CA) up to 2-3 yrs	Consider further assessment of intake and medical conditions that may impact growth if growth is less than expected: • Weight/length <5th percentile • No weight gain • Weight loss • Decline in growth across channels	Rate of linear growth: • 0.5-1.0 cm/week up to 6 months of age • May decline 6-12 months, compare to expected rates for term infants using CA Note: Infants who do not demonstrate equivalent decreases in growth rate after 6 months of age may demonstrate "catch-up growth." Rate of weight gain: • 20-30 g/day up to 6 months of age • May decline 6-12 months, compare to expected rates for term infants using CA

continued...

Assessment (continued)	Intervention	Evaluation/Outcome
Dietary		
Assess adequacy of fluid intake for age and size.	If fluid intake not adequate to meet fluid needs: • Check for feeding difficulties • Consider changes in feeding frequency, volume, position or environment	Infant is well hydrated.
Assess adequacy of energy intake. Assess adequacy of formula volume for energy needs.	If feeding difficulties and/or growth concerns, but fluid intake is adequate, consider increasing energy density of formula.	On 20 kcal/oz formula, 2.75 oz of formula/lb of infant weight (180-190 cc/kg/d) provides 120 kcal/kg/day.
Assessment (continued)	**Intervention**	**Evaluation/Outcome**
Assess appropriateness of vitamin/ mineral intake.	When intake is <24 oz, provide multivitamin supplement. Breast/ chestfed infants should continue receiving a vitamin supplement for Vitamin D and Iron. Provide 2-4 mg/kg iron (in formula or as supplement) for first year or until 12 months CA.	Infant receives appropriate amounts of vitamins and minerals.
Check appropriateness of type of feeding.	Recommend appropriate type of feeding.	Breast/chest milk or standard infant formula to 12 months CA. (Soy formula is not recommended for premature infants at risk for osteopenia.) If on non-standard types of feeding, evaluate energy, protein, fluid, and vitamin/ mineral intake. Assure that intake is consistent with recommendations and individual infant condition and needs.
Check appropriateness of feeding transitions for developmental age (use CA).	For infants with history of growth or other nutrient deficiencies, consider selection of transitional foods that will meet specific needs of infant. For infants having difficulty making feeding transitions: • Evaluate developmental readiness • Evaluate that feeding relationship is supportive of infant skill and development • Consider referral to appropriate specialty for evaluation	Infant shows progress in feeding related to appropriate developmental level.

[1] For reference data and guidelines for taking accurate measurements, see Chapter 2.

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Chapter 15

NUTRITION INTERVENTIONS FOR RESPIRATORY DISEASES

Donna Johnson, RD, PhD and Joan Zerzan, MS, RD, CD
Updated by Mari Mazon, MS, RDN, CD and Merideth Murray, MS, RDN

Causes of abnormal respiratory function in infancy are shown in Table 15-1. Nutritional implications depend on the nature of the disorder, clinical manifestations, treatment modalities, compensatory mechanisms, and the presence of other underlying conditions. Compensatory mechanisms are designed to maintain normal ventilation and include increased respiratory rate, increased work of breathing, grunting, and nasal flaring. The consequences of these mechanisms include increased effort, increased energy expenditure, and exhaustion. Normal infant activities such as feeding and growth may be impacted.

Table 15-1: Causes of Respiratory Disease in Infancy (1)

Cause	Example
Infections	Viral (respiratory syncytial virus-RSV, parainfluenza), bacterial
Bronchopulmonary dysplasia (BPD), Chronic Lung Disease (CLD)	prematurity, aspiration, meconium aspiration, infection
Congenital anomalies of heart and lung	Tracheomalacia, laryngomalacia, congenital heart disease (CHD), diaphragmatic hernia, hypoplastic lung, congenital cysts or tumors
Congenital Syndromes	Beckwith-Wiedeman, CHARGE association, Pierre Robin, Treacher Collins, Trisomy 18, deLange, Mobius sequence, cleft lip and palate

Bronchopulmonary dysplasia (BPD) is the most common form of chronic lung disease seen in preterm infants (2). It was first described in 1967 as a lung injury resulting from mechanical ventilation and aggressive oxygen support (3.4) It is now known that these interventions disrupt lung alveolarization and vascularization, leading to impaired lung development (4).

Advances in respiratory care such as the use of gentler ventilation, antenatal steroids, and postnatal surfactants have reduced the severity of BPD, particularly in infants born weighing more than 1200 g or after 30 weeks gestation (4). However, the incidence of BPD has remained largely unchanged in infants born at or before 28 weeks gestational age (2).

Diagnostic criteria for BPD have evolved overtime. The latest definition was developed in 2001 under the collaboration of the National Institute of Child Health and Human Development

(NICHD), National Heart, Lung and Blood Institute (NHLBI), and Office of Rare Diseases (ORD). It is described below.

Gestational Age	<32 weeks	≥ 32 weeks
Time point of assessment	36 weeks PMA[a] or discharge to home, whichever comes first	>28 days but <56 days postnatal age or discharge to home, whichever comes first
Treatment with oxygen >21% for at least 28 days **plus**		
Mild BPD	Breathing room air at 36 weeks PMA[a] or discharge, whichever comes first	Breathing room air by 56 days postnatal age or discharge, whichever comes first
Moderate BPD	Need for <30% oxygen at 36 weeks PMA[a] or discharge, whichever comes first	Need for <30% oxygen at 56 days postnatal age or discharge, whichever comes first
Severe BPD	Need for ≥30% oxygen and/or positive pressure, (PPV[b] or NCPAP[c]) at 36 weeks PMA[a] or discharge, whichever comes first	Need for ≥30% oxygen and/or positive pressure (PPV[b] or NCPAP[c]) at 56 days postnatal age or discharge, whichever comes first

[a] Post menstrual age
[b] Positive-pressure ventilation
[c] Nasal continuous positive airway pressure

This chapter will cover concerns that are specific to the infant with chronic pulmonary dysfunction (BPD/CLD).

Growth and Nutritional Support

The overall goal for infants with BPD/CLD is to promote growth and development. As infants grow, lung function improves and risk of severe cardiopulmonary sequelae, morbidity, and mortality with respiratory infection declines. Cornerstones of treatment are pulmonary support to maintain optimal oxygen saturation and prevent complications and nutrition support to promote growth.

Growth

Growth in infants and young children with BPD may be compromised by several factors. These include respiratory limitations, altered nutrient needs, drug-nutrient interactions, and feeding difficulties. Studies of growth in patients with BPD/CLD show reduced percentile ranking and reduced fat-free mass and total body fat during the first year of life (5). Although long-term follow-up suggests that difference in growth continues into school age, with children who had BPD as an infant measuring shorter and lighter than peers who were born full-term without BPD, such studies must be interpreted with caution as the treatment these children received as infants may have followed outdated guidelines (6).

Persistent hypoxemia is recognized as a cause of poor growth as well as feeding problems in children with BPD (5, 7). Inappropriate discontinuation of oxygen therapy for these children has been reported to cause an abrupt drop in growth rates. Maintaining adequate oxygenation

improves growth (8). Infants with BPD who are not on oxygen therapy may experience oxygen desaturation with feeding after hospital discharge (9). Oxygen saturation should be assessed when growth falters or when fatigue and aversive behaviors are observed during feeding.

Medications used in the management of BPD/CLD may contribute to feeding intolerance, nutrient utilization, or nutrient needs. Through these interactions, growth may be negatively impacted. Medications frequently used in the treatment and management of BPD/CLD include diuretics, steroids, methylxanthines, and bronchodialators. Many oral medications are hyperosmolar and when added to feedings can contribute to nausea and feeding intolerance. Some medications may increase metabolic rates and result in increased energy needs. Long-term use of steroids negatively impact linear growth and alter mineral status. Diuretics increase urinary losses of electrolytes, calcium, magnesium, and zinc. A thorough history should include medications and identify specific monitoring needs (4, 7).

Growth faltering due to inadequate intake in infants with BPD/CLD has been documented. Factors contributing to inadequate intake include feeding difficulties, recurrent illness, and increased energy needs (4).

Nutritional Support

Nutrition is critical for prevention, treatment, and recovery from BPD/CLD. Antioxidant nutritional therapies, including Vitamin A, Vitamin E, selenium and n-acetylcysteine, have been proposed for the prevention of BPD (7). It has also been proposed that inositol may aid in the prevention of BPD by enhancing the production of surfactant (10). These therapies require further investigation. Recovery from BPD occurs with growth of new lung tissue. Supportive therapy, therefore, depends on the provision of adequate nutrients to support growth (10, 11).

Higher energy needs have been proposed as a cause of growth failure in infants with BPD/CLD. It has been suggested that infants with respiratory dysfunction may experience increased energy expenditure associated with increased work of breathing (WOB). Studies, however, have not demonstrated this consistently (10). Some infants with BPD experience increased energy needs (12, 13). The reasons for this are not entirely clear, but increased work of breathing, catecholamine release due to stress, increased energy requirements for feeding, and the effects of medications probably all play roles. Energy requirements of 140-150 kcal/kg/d have been proposed for infants with BPD based on an estimated 15-25% greater need compared to healthy growing preterm infants (14). A number of factors contribute to energy expenditure in individuals, including genetics, activity, and severity of respiratory distress. Correlating growth with energy intake is the best indicator of adequacy.

It may be difficult to provide adequate energy to infants and young children with BPD. They may have ongoing fluid restrictions due to concerns about pulmonary edema. They may experience

fatigue with feeding. Increasing the energy density of formula or breastmilk using a combination of components may be helpful. For infants with BPD it is inappropriate to use only carbohydrate to increase energy density. A high carbohydrate load increases production of CO2. At the same time, the addition of excess fat may delay gastric emptying. Delayed gastric emptying may contribute to gastroesophageal reflux. The addition of vegetable oils that may separate out from formula or breastmilk may be problematic as they may increase the risk of aspiration pneumonia. Since infants with BPD are at risk for more frequent and serious illnesses in the first months of life, it is important to teach caregivers how to assess hydration status during illness, especially when infants are receiving an energy-dense formula.

Nutritional care for the infant with BPD must be individualized. Feeding concerns, nutrient needs, and growth outcomes are different for each infant. Variables that influence the nutrition care plan include initial severity of BPD, presence of other medical problems, and characteristics the infant and caregiver bring to the feeding relationship. Infants and young children with severe BPD may require ongoing mechanical ventilation and a tracheostomy, medications with nutrition implications such as corticosteroids and diuretics (see Chapter 5), gastrostomy tube feedings (see Chapter 10), and frequent hospitalization. Some infants with milder forms of BPD may continue to require medical/nutritional interventions post discharge including supplemental oxygen, medications, and tube feeding. Some infants with CLD may experience exacerbations of respiratory dysfunction with illness, and have feeding difficulties without ongoing need for oxygen, medications, or tube feeding.

Feeding Difficulties

Feeding problems are common among infants with moderate or severe BPD. These infants benefit from an interdisciplinary team approach to assess and treat feeding issues. Problems found in infants with BPD include poor coordination of suck, swallow, and breathing, swallowing dysfunction with silent microaspiration, oral-tactile hypersensitivity, and aversive behavior associated with unpleasant oral and feeding experiences. Assessment of feeding problems with feeding observations, swallowing studies, and measurements of oxygen saturation during feeding may be helpful. Infants with BPD may also experience gastroesophageal reflux and/or delayed gastric emptying. Feeding may show significant improvement if these conditions are diagnosed and treated with changes in feeding patterns, positioning, or medications (15,16,17) (See Chapter 8).

The growth and development of infants with BPD is also influenced by family characteristics. Taking care of these infants can present many challenges. Feeding issues may contribute to the stress of caring for an infant with BPD. Feeding infants with moderate and severe BPD may require several hours each day (15,18). Nighttime feedings may last for several months. Health care professionals and the families themselves may put excessive emphasis on weight gain increments and establish problematic feeding behavior patterns. Infants with BPD are often

rehospitalized. They are at high risk of serious illness during the respiratory syncytial virus (RSV) season from November through March, and families are usually told to keep their babies at home. Many caregivers report a sense of social isolation. Assuring that family needs for social, emotional and financial support are met is an essential component of good care for these infants and young children.

The remainder of this chapter presents guidelines for nutritional assessment, intervention, and evaluation/outcome for children with broncopulmonary dysplasia.

Table 15-1: Nutrition Interventions for Bronchopulmonary Dysplasia

Screening/Assessment	Intervention	Evaluation/Outcome
Anthropometric		
Measure and plot on appropriate growth chart using corrected age:*† • Weight for age • Length for age • Weight for length or BMI • Head circumference (under 2 years) During hospitalization, daily weights and weekly length measures are recommended for monitoring growth and fluid status.[4] After discharge, frequency of monitoring will depend on the child's growth and presence of feeding challenges. Growth goals are similar to reference rates for corrected age. Evaluation of Z-scores and growth velocity can help identify infants and children with malnutrition.§ Infants relying on tube feeding should have frequent weight checks to adjust feeding rates with growth, initially every 1-2 weeks, then monthly. While some infants and young children with BPD will exhibit catch-up growth (growth that shows increasing percentiles on growth charts), those who were very small at birth and those who have moderate to severe illness may not show catch-up growth for several years. All infants and children with BPD should have at least a steady growth pattern that follows established growth percentiles.	Further assessment is required to determine potential causes of inadequate growth and malnutrition status. Intervention may be a combination of dietary, feeding, behavioral and medical interventions. If long-term linear growth is poor, assess protein intakes.	Maintain established growth pattern for weight and length for age.
For older toddlers and children with moderate to severe BPD, especially those on corticosteroid therapy, assessment of mid-arm circumference, mid-arm muscle circumference, and triceps skinfold can be useful to assess fat stores, protein status, and presence of malnutrition.	Very inactive children and those dependent on steroids or mechanical ventilation may develop excessive fat stores and energy intake may need to be reduced.	Fat and muscle stores within normal parameters for age, gender, and medical condition.
Biochemical See laboratory standards for normal values		
Iron Status: Measure complete blood count if iron deficiency is suspected[4] (eg. Inadequate iron intakes).	If lab values indicate possible iron deficiency anemia, assess dietary and supplemental iron intake and apply dietary methods to increase iron intake and absorption and/or consider trial dose of increased iron supplement.	Indicators of iron status are within normal limits.

Bone mineralization: For infants at high risk of osteopenia of prematurity (those on long-term diuretics, corticosteroid therapy, or parenteral nutrition[11] and those fed unfortified human milk, term formula, or soy formula before achieving weights of 2000 gm) measure alkaline phosphatase, calcium, phosphorus, vitamin D, and parathyroid hormone[4].	If lab values indicate risk of osteopenia: • consider supplementing with calcium, phosphorus, and vitamin D • discuss possibility of changing corticosteroid dosing patterns (e.g., every other day) or methods of delivery (e.g., inhaled vs. systemic) with primary care provider	Indicators of bone mineralization are within normal limits.
Electrolyte balance: For infants and children on diuretics measure electrolytes (sodium, chloride, and potassium), calcium, zinc, magnesium[4]	• Discuss with primary care provider about supplementing with electrolytes or minerals. Discuss possibility of changing type or dose of diuretics with primary care provider.	Indicators of mineral and electrolyte status are within normal limits.
Clinical		
Oxygen status: Periods of hypoxemia or marginal hypoxemia should be suspected whenever infants with BPD fail to grow.[1,18,20] Previously undetected hypoxemia has been reported during sleep and during and after feeding.[21] Oxygen status should be assessed with pulse oxymetry during feeding, sleeping, and crying. SaO2 >92% during feeding, sleeping, and crying is recommended.[1] Values in the range of 95% have been reported to increase growth.[1]	Provide oxygen therapy as needed. This may include oxygen support only at feeding and sleeping or increased flow rates at these times.	Growth is appropriate. SaO2 remains >92%.[11]
Fluid restriction: Fluid restriction may be prescribed for infants with severe BPD in first months of life.	Plan diet that provides adequate energy and nutrients with limited fluid intake. May need to concentrate formula.	Growth is appropriate.
Gastroesophageal reflux (GER): Assess presence of GER symptoms: regurgitation with gagging/coughing/repeated swallowing between meals, red and teary eyes, excessive vomiting, esophagitis (post prandial pain, anemia), respiratory symptoms (pneumonia, wheezing), neurobehavioral symptoms (irritability, crying, feeding refusal, seizure-like attack).	If symptoms indicate need for further assessment, refer to primary care provider for arrangements for diagnostic tests. Interventions for GER include: • dietary (e.g. smaller frequent feedings) • positioning • thickening (for infants >42 weeks gestation) • medication • surgical (reflux unresponsive to above treatments may require surgical intervention)	Meal times and post-prandial period are pleasant and pain free. Respiratory symptoms improve.

continued...

Screening/Assessment (continued)	Intervention	Evaluation/Outcome
Dietary		
For infants: assess intake of energy, protein, carbohydrate, vitamins, minerals. In infancy, protein intake may be compromised if intake of infant cereal, high carbohydrate baby food, or glucose polymers is excessive. Protein should provide 8-12% of energy for infants. Excessive protein and high renal solute load may result if formula is concentrated to greater than 24 kcal/oz without the use of modular products.	For infants with mild BPD, appropriate growth, and good feeding skills, provide standard infant formula. For infants with moderate or severe BPD, ongoing fluid restriction, or feeding problems that interfere with adequate intakes, a 22 kcal/oz formula may be used or formula may be concentrated to 24-30 kcal/oz. Guidelines for infants receiving tube feedings are found in Chapter 10. Small infants with limited energy needs may require additional supplements to meet DRI/RDA requirements for protein, vitamins and minerals if sufficient formula is not taken. Attention should be paid to iron, calcium and phosphorous intakes of all VLBW infants (see Chapter 14).	Growth is adequate. Intakes of vitamins and minerals are at DRI/RDA levels.
Assess introduction of non-milk feedings (solids). Foods are sometimes introduced to the infant with BPD according to chronological age since birth. This is not nutritionally or developmentally appropriate. Infants with growth and feeding difficulties should begin the transition to solid foods using high nutrient dense foods. This can be accomplished by reading labels for jar baby foods, using infant cereal mixed with breast milk or formula, and when appropriate making baby food from table foods.	Foods should be introduced as the infant is developmentally ready. See Chapter 8.	Foods are introduced to the child when developmentally appropriate.

Assess intake of energy, protein, vitamins, and minerals of young children.	If energy needs remain high past early infancy, foods should be chosen to provide optimal energy and nutrients. Suggestions include yogurt, pudding, cottage cheese, pancakes, hot cereals, tuna or meat salad, scrambled egg, cheese, and mashed avocado. High fat foods such as butter, margarine, mayonnaise, cream cheese, and cream can be added to other foods to increase energy content. Homemade milkshakes, fruit slushies, and instant breakfast products can be used as an energy-dense snack or bedtime beverage. Commercial pediatric enteral feeding products may also be used for this purpose. Meals and snacks should be offered at regular times 5-6 times each day in a pleasant, non-coercive environment. Tube feedings are sometimes required for infants with BPD, especially those who continue to depend on mechanical ventilators. The type of enteral feeding is usually changed at about 1 year of age. (See Chapter 10). Tube-fed infants should continue to receive oral stimulation and to have social interactions at feeding times.	Young children are growing adequately and achieving DRI/RDA levels of vitamin and mineral intake.
Feeding skills: Feeding problems are common in infants with BPD. Feeding should be assessed by a feeding observation and careful questioning of caregivers.	A team approach to feeding problems and referral for additional assessment and therapy may be indicated. (See Chapter 8)	Problems with feeding are addressed. Child ingests adequate energy to support growth. Family is able to enjoy feeding interactions with child.

continued...

Screening/Assessment (continued)	Intervention	Evaluation/Outcome
Fatigue: Consider fatigue as a feeding issue if the infant stops feeding before ingesting adequate energy.	Check with primary care provider about provision of additional oxygen at feeding times and/or use of bronchodilators before feedings. Increase energy concentration of formula or other foods. Manipulate the feeding schedule to increase efficiency. Shorten feeding times and end when feeding becomes less efficient. Try smaller, more frequent feedings. For older infants, make sure food textures offered match infant's oral-motor skills. Consider gastrostomy tube placement. (See Chapter 10)	Child ingests adequate energy to support growth.
Poor coordination of swallowing: Feeding observation may show abnormal sucking patterns with short irregular sucking bursts with long pauses and rapid breathing.	See Chapter 8. Help infant to "pace" feeding. Consider interventions listed in fatigue section (above). Refer for feeding therapy.	Infant feeds without distress and demonstrates coordination of suck-swallow-breathe.
Swallowing dysfunction due to aspiration: Infants with BPD are at risk of aspiration due to airway damage caused by intubation as well as reduced ability to use pulmonary air to clear the larynx. Suspect aspiration with episodes of respiratory deterioration or wheezing with feedings, and refer for testing and intervention. Aspiration can be assessed with videofluoroscopic swallowing study (VFSS)	Dependent on findings of specialist. May include changes in texture or temperature of foods and beverages. In severe cases it may not be safe to feed orally.	Feeding is not associated with adverse pulmonary consequences.

Oral-tactile hypersensitivity: Infant becomes agitated, pulls back, gags, or vomits when oral feeding is attempted. Infants and young children with BPD are at increased risk due to aversive oral experiences early in life. This situation often requires the intervention of specialists in pediatric feeding therapies.	See Chapter 8. Refer to feeding therapy for de-sensitization. Avoid aversive oral experiences as much as possible. Gradually introduce pleasant oral-tactile experiences into daily care routines. Encourage oral exploration.	Child displays pleasure with feeding and oral exploration.
Behaviors: Infants and children with BPD are at risk for developing inappropriate feeding behaviors and interactions.	See Chapter 9.	Problems with feeding behaviors are addressed.

* For reference data and guidelines for taking accurate measurements, see Chapter 2.
† For information about correcting for prematurity, see Chapter 14.
§ For information about identifying malnutrition, see Chapters 1 and 2.

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Chapter 16

NUTRITION INTERVENTIONS FOR CYSTIC FIBROSIS

Ellen Bowser, MS, RDN, LDN, RN, FAND, Valerie Eubanks Tarn, MS, RDN, LDN and Umme Salma Vahanvaty, MS, RDN

Introduction:

Cystic fibrosis (CF) is an autosomal recessive genetic disorder caused by a mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene. It is a rare, progressive, life-limiting disease that affects all ethnicities.

The CFTR gene (NM 000492) and mutations identified with the syndrome of CF were identified in 1989(1). The most common CFTR mutation is F508del; 85.4 percent of individuals in the Cystic Fibrosis Foundation (CFF) Patient Registry who have been genotyped have at least one copy of this mutation. To date, more than 2,000 mutations have been found in the CFTR gene (2,3).

According to the 2023 CFF Patient Registry Annual Data Report, there are 33,288 people living with CF in the United States. The prevalence of CF in the US is approximately 1 in 3,900. Among people with CF born between 2019 and 2023 half are predicted to live to more than 61 years old (3). The number of adults with CF keeps increasing, while the number of children with CF has remained stable.

The prevalence of CF in the US is highest among the white population.

Role of CFTR gene

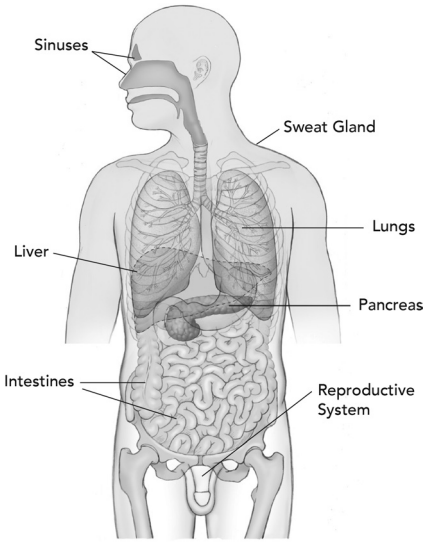
The CFTR protein is a plasma membrane bound cyclic-AMP sensitive anion channel that is involved in the transport of chloride ions to the surface of the cell and intake of sodium ions into the cell.

When the protein is not working properly, it is unable to move chloride ions to the epithelial surface where it draws water in from outside the cell. Without this, the mucus on the cell surface becomes dehydrated, thick and sticky. It does not allow the cilia on the cell surface to effectively move the mucus across the surface of the epithelium.

In the lungs, the thick and sticky mucus traps bacteria and other germs leading to infections, inflammation, and obstruction of the airways. In the pancreas, the buildup of mucus does not

allow the adequate release of enzymes required for absorption of nutrients leading to weight loss, poor growth, and micronutrient deficiencies particularly fat-soluble vitamins. In the liver the thick mucus can block the bile duct, causing liver disease. In persons with male anatomy, it can cause blockage of the vas deferens, thereby affecting fertility (see Figure 16-1) (4).

Figure 16-1: Cystic Fibrosis's Effects throughout the Body



Cystic fibrosis causes changes in many parts of the body, including the lungs, pancreas, liver, intestines, sinuses, reproductive system, and sweat glands.

From CFF.org (5)

Diagnosis of CF

Cystic fibrosis causes changes in many parts of the body, including the lungs, pancreas, liver, intestines, sinuses, reproductive system, and sweat glands.

Early diagnosis of CF is possible as part of the universal newborn screening program (NBS) and prenatal testing. Some people are diagnosed as adults. After a positive newborn screen, the infant is referred to an accredited CF center for further work-up. This involves genetic testing and a sweat test. All infants or persons with CF (PwCF) also need to complete a stool or fecal elastase test to determine if they are pancreatic sufficient (PS) or insufficient (PI).

All PwCF should be followed by an interprofessional team at an accredited CF Center. The clinical practice guidelines published by the CFF require a coordinated, interprofessional approach for the management of PwCF who have diverse needs and complex treatment plans (6). Typically, the treatment involves professionals in the following areas:

- Medicine
- Nursing

- Nutrition
- Physical therapy
- Respiratory therapy
- Social service
- Pharmacy
- Psychology
- Gastroenterology
- Endocrinology
- Infectious disease
- Otolaryngology

Nutritional Assessment

Nutritional assessment for children with CF includes anthropometric measurements, dietary history, pancreatic enzyme assessment, and stooling history. Anthropometric measurements including height/length, weight, and BMI should be obtained at each visit (see Chapter 2). Growth parameters should be plotted on appropriate growth charts based on sex assigned at birth and age. The CFF recommends children less than 2 years of age have their weight, length and weight for length plotted on the WHO growth charts. Children ages 2 to 20 should have their growth measures plotted on the CDC growth charts (6). Growth goals for children less than 2 years of age are weight/length at the 50th percentile or greater, and the goal for those 2 years and older is a BMI at the 50th percentile or greater. These goals are recommended as this level of growth is associated with near normal lung function in children with CF. Weight gain in children with CF should be similar to their peers without CF (7).

Dietary history may include a 24-hour recall, timing of meals and snacks, and the use of oral supplements and multivitamins (see Chapter 1). The history should identify eating behaviors such as extreme pickiness, food jags, and length and location of meals. Additionally, the timing of pancreatic enzyme supplementation and how they are administered should be determined. Stooling history, which includes the number of stools per day, abdominal discomfort, bloating and/or gas, should be included in the nutritional assessment of the child with CF. Fat-soluble vitamin levels and other biochemical indices are assessed annually by the CF Center staff. Center staff should also ask families to identify their participation in programs such as WIC and SNAP food benefits.

See Figure 16-2 at the end of the chapter for an example nutrition screening form for PwCF. It is included as context for community RDNs who concurrently see PwCF that are managed at an accredited CF Center. To improve nutrition outcomes and avoid duplication of services, it is important for the community RDN and CF Center RDN to partner together and coordinate care.

Enzyme Administration

The majority (85-95%) of children with CF will have exocrine pancreatic insufficiency (PI). PI presents shortly after birth or within the first year of life (8). Signs of PI include large, frequent, bulky stools, gas, and bloating, which can lead to poor weight gain, weight loss, and fat-soluble vitamin deficiencies. A stool or fecal elastase test is used to diagnose PI.

PI is treated with oral pancreatic enzyme replacement. Pancreatic enzymes contain lipases, proteases, and amylases. There are several brands and doses of these products approved by the Food and Drug Administration (FDA). Generic enzymes are not recommended. Oral enzymes are microspheres coated with an acid-resistant formulation and packaged in capsules.

Dosing with oral pancreatic enzymes is based on the units of lipase per kilogram (kg) body weight per meal. Recommendations from the CFF are to not exceed 2,500 units of lipase per kg body weight per feeding or meal or greater than 4,000 units of lipase per gram of fat. The total daily dose should not exceed 10,000 units of lipase per kg body weight per day (8).

The capsules can be swallowed whole or opened up and the beads sprinkled on a small amount of acidic food. Enzymes should be taken immediately prior to all meals and snacks, including human milk. Generally, the snack dose is half the dose given with meals. PwCF and their families should be taught how to administer enzymes and how to adjust capsule number based on the size and fat content of the meals and snacks. Tube feedings also require pancreatic enzyme supplementation.

Vitamins and Minerals

PwCF who are pancreatic insufficient (PI) are at greatest risk of vitamin deficiencies due to malabsorption. They are required to take CF-specific multivitamin and mineral formulations that are designed for better absorption. There are 6 different brands of multivitamins to choose from: MVW Complete Formulation® (original and with additional Vitamin D- 3000IU OR 5000IU), AquADEKs, DEKA Plus, GenADEK®. See Table 16-2 at the end of the chapter for a comparison of the micronutrient composition of these multivitamins (9).

All of the CF-specific multivitamins contain vitamins A (as beta carotene), E, D, K, B-vitamins, and zinc. They do not contain calcium or iron, and some PwCF may need additional supplements for these minerals.

PwCF have higher sodium needs due to the loss of sodium in their sweat. They lose 2-3 times more salt in their sweat as compared to people without CF. Additional salt needs to be provided from birth (see Table 16-1). It is usually added to the breast/chest milk or formula, or it is given as a salt solution.

Table 16-1: Infant Salt Dosing Guidelines

Age	Daily Salt Dosage
Birth-6 months	1/8 teaspoon (13 mEq)
>6 months, as long as the infant is on the growth curve for a six month old infant (10)	1/4 teaspoon (26 mEq)
Salt dosage is given in small increments throughout the day. *After 12 months of age, it is recommended that toddlers include salt in their foods.*	

While physically active or in hot weather when there is an increased loss of salt in sweat, PwCF are advised to increase their salt intake by adding salt to electrolyte-rich drinks or consuming salty snacks, like pickles, beef jerky, cheese, or pretzels.

Adequate fluids along with high salt intake are both required to prevent dehydration in PwCF, especially in the summer months when outdoors or exercising (11, 12).

Oral Supplements and Tube feedings

The nutrient needs of PwCF are often difficult to meet because of both increased nutrient requirements and decreased food intake. The energy requirement is increased because of hypermetabolism intrinsic to the genetic defect, increased losses of nutrients attributable to pancreatic insufficiency, chronic pulmonary infection, and sometimes sinusitis. Decreased intake is related to CF exacerbation, anorexia from both GI and pulmonary symptoms, and psychosocial issues, such as the deliberate restriction of foods that may be perceived as “bad” foods and the cost and labor involved in preparing meals.

PwCF may need to boost their caloric intake during specific growth periods. Families are encouraged to utilize the below strategies to meet their caloric needs.
Calories are increased in the following ways:

1) Consume 3 meals and 3 snacks daily. Portion sizes should be larger than age appropriate.
2) Calorie boosters, like additional fat and protein, may be added to foods and beverages. The most common calorie boosters are oil, butter, cheese, cream, sour cream, nuts, nut butters, dried fruits, and avocados.
3) High calorie oral supplements (homemade or commercially available) may be necessary. The cost of some of these may be covered by various patient assistance programs, or insurance company with a medical justification.
4) A feeding tube may be used to supplement caloric intake if the previous options and the use of appetite stimulants have failed to result in weight gain and improvement in BMI for age. For most PwCF, feeding tubes are used to provide ready to feed, commercially available formulas overnight. During the daytime, PwCF will continue to eat by mouth. The goal is to meet at least 50% of caloric needs from the feeding tube.

Appetite Stimulants

Appetite stimulants should be used only if the etiology of inadequate intake is identified as a poor appetite without underlying GI or psychological causes.

Appetite stimulants are usually prescribed for short-term use to boost intake and promote weight gain. Based on the age of the PwCF, different appetite stimulants will be prescribed by the physician. The most common appetite stimulants are Cyproheptadine (Periactin®), Dronabinol (Marinol®), Megestrol (Megace®), and Mirtazapine (Remeron®). All appetite stimulants have side effects that the physician will discuss with the family or PwCF before prescribing.

Comorbidities

Other related comorbidities in CF are more common due to the increased median age of survival. Some individuals develop CF-related diabetes (CFRD) as the incidence and prevalence of this condition increases after age 10 (11). The 2022 CF Patient Registry shows 4.5% of those < 18 years and 29.7% of those > 18 years were diagnosed with CFRD (2). Symptoms related to CFRD include difficulty gaining weight, decreased pulmonary function, or increased pulmonary exacerbations. In contrast, the classic symptoms of type 1 and type 2 diabetes include frequent urination, increased thirst, and increased hunger.

The oral glucose tolerance test (OGTT) is the gold standard for diagnosing CFRD and is recommended annually by age 10 or earlier if symptoms occur (11). Insulin replacement therapy is the only treatment for CFRD, and the type and amount of insulin should be determined by an endocrinologist. Individuals should continue to eat a healthy diet that is high in calories and protein. Sweetened drinks are discouraged because they can exacerbate high blood sugar. The goal in CFRD is to achieve and maintain optimal nutrition status and normalize blood sugars.

CF-related bone disease (CFBD) is another complication seen in PwCF. Contributing factors for poor bone health include low body mass index (BMI <25th percentile), severe lung disease (FEV1 <50th percentile predicted), chronic steroid use of >/= 5mg/day for >/= 90 days/yr, delayed puberty, or a history of fractures. It is recommended to obtain a dual-energy x-ray absorptiometry (DXA) scan on all adults >18 years old and children > 8 years old if they have any of the above-mentioned risk factors (14). To optimize bone health, it is important to achieve vitamin 25 (OH) D level of at least 30ng/mL, consume adequate calcium, participate in weight bearing exercise, and maintain a BMI in the 50-85th percentile.

Highly Effective Modulator Therapies

The FDA approved the first drug targeting the underlying cause of CF over a decade ago. Today, CFTR modulator therapy is available for approximately 90% of PwCF. Modulators are mutation-specific, and there are currently four approved drugs: Kalydeco® (ivacaftor), Orkambi® (lumacaftor/

ivacaftor), Symdeko®(tezacaftor/ivacaftor), and Trikafta® (elexacaftor/tezacaftor/ivacaftor). Some of these treatments have shown remarkable improvements in lung function and growth. The effect of CFTR modulator therapy on anthropometric parameters depends on the genetic mutation and the type of modulator therapy used (15). Most recently, there has been an increase in overweight/obesity in people with CF due to improved care. The emphasis on a healthy diet versus the traditional high calorie, high fat CF diet should be addressed in this population. Overall, nutrition care should be individualized using clinical data and the goals of the PwCF (16).

Figure 16-2: CF NUTRITION SCREENING FORM

Insurance or other nutrition program: (Medicaid, WIC)_____

Anthropometrics:

Wt (kg): _____,_____%; _____z-score Wt/Ht:_____,_____%; _____z-score

Ht (cm): _____,_____%; _____z-score

BMI(kg/m2): _____,_____%; _____z-score

Wt GAIN/ LOSS _____x_____ days (average_____grams per day)

Enzymes/Medications:

Brand: _____ Capsules per meal: _____Capsules per snack: _____

Total units lipase/kg/meal: _____ Total units lipase/kg/day _____

Other Nutrition Related Medications: (ex.CFTR modulators, insulin, CF vitamins, growth hormone, proton pump inhibitor etc.)

_____

Estimated Nutritional Needs:

EER: _____

Energy: kcal/kg: _____ Protein: g/kg: _____

Fluids mL/day: _____

What oral high calorie nutrition supplements do you use? _____

If so, how many per day? _____

Do you check Blood Glucose Levels? YES/ NO

If so, what did they range last week? _____

Do you exercise? YES /NO

If so, what type of exercise? _____

How is your appetite?	Great	Good	Fair	Poor	
What is your stool like?	Formed	Loose	Oily	Hard	Bulky
How many stools per day?	<1	1	2	3	>3
GI Symptoms?	Pain	Distention	Nausea	Gas	Constipation
	Vomiting	Reflux	Bleeding		

Anthropometrics

- Birth to 2 years of age, WHO growth chart, goal wt/length 50th percentile or greater
- 2 to 20 years, CDC growth chart, goal BMI 50th percentile or greater
- Growth Chart tool for all ages: www.peditools.org

Enzyme Replacement Therapy

- Take lipase units x number of capsules at meals /weight (kg) = units of lipase/kg/meal
- Take lipase units x number of capsules per day/weight (kg) = units of lipase/kg/day
- Enzymes should be given before meals and snacks
- Enzyme dosage should be in the range of 1,500 to 2,500 units of lipase/kg/meal and not exceed 10,000 units of lipase per kg/day
- Ex: Creon 24,000® has 24,000 units of lipase per capsule
- Enzyme dose: Creon 24,000®, 4 capsules with meals and 2 with snacks

  Weight: 51kg,
  Creon 24,000® x 4 capsules/meal = 96,000 units lipase/meal
  96,000 units lipase/meal divided by 51 kg = 1882 units lipase/kg/meal

Estimated Energy Needs

- Energy: EER for age x CF factor of 1.2-2.0
- Protein: DRI x CF factor of 1.5-2.0
- Fluid: Weight < 10kg = 100mL/kg ; Weight >10kg = 1500-1800mL/m2

Sample Nutrition Diagnosis:

Problem 1: Altered GI function
Related to: CF with pancreatic insufficiency
As evidenced by: patient requiring pancreatic enzyme therapy for food digestion/absorption

Problem 2: Increased energy needs
Related to: Increased work of breathing and malabsorption
As evidenced by: the needs for high calorie, high protein diet, GT feeds nightly, and pancreatic enzyme therapy to promote optimal nutrition

Table 16-2: Vitamin Comparison Chart for CF-Specific Multivitamins (Updated April 2021 – Used with Permission)

FAT-SOLUBLE VITAMINS & ZINC[1]								
CFF SPECIFIC VITAMINS				GUIDELINES FOR VITAMIN D MANAGEMENT[3]				
				STEP 1			STEP 2	
MVW Complete Formulation® Drops Chewables, ADEK Gummies[6] Softgels / Softgel Minis	**GenADEK®** Drops Chewables (Q3 2021)	**AquADEKs** Drops Chewables	**DEKAs Plus** Drops Chewables	**MVW Complete Formulation®** D3000 Chewables D3000 Softgels	**GenADEK® STEP 1** Softgels	**DEKAs Plus** Softgels	**MVW Complete Formulation®** D5000 Chewables D5000 Softgels	**GenADEK® STEP 2** Softgels
Total Vitamin A IU / mcg Retinol Activity Equivalents (RAE) (Retinol and Beta Carotene)								
4,627 IU / 1,388 mcg RAE / 0.5 ml 75% as beta carotene	5,768 IU / 1730 mcg RAE / 1 ml 87% as beta carotene	5,751 IU / 1,725 mcg RAE/ 1 ml 87% as beta carotene	5,751 IU / 1,725 mcg RAE/ 1 ml 87% as beta carotene	*NP*	*NP*	*NP*	*NP*	*NP*
9,254 IU / 2,777 mcg RAE / 1 ml 75% as beta carotene	11,536 IU / 3,460 mcg RAE / 1 ml 87% as beta carotene	11,502 IU / 3,451 mcg RAE/ 2 ml 87% as beta carotene	11,502 IU / 3,451 mcg RAE / 2 ml 87% as beta carotene	*NP*	*NP*	*NP*	*NP*	*NP*
16,000 IU / 4,800 mcg RAE / 1 chewable 88% as beta carotene	18,167 IU/ 5,450 mcg RAE / 1 chewable 92% as beta carotene	18,167 IU / 5,450 mcg RAE / 2 chewables 92% as beta carotene	18,167 IU / 5,450 mcg RAE / 1 chewable 92% as beta carotene	16,000 IU / 4,800 mcg RAE / 1 chewable 88% as beta carotene	*NP*	*NP*	16,000 IU / 4,800 mcg RAE / 1 chewable 88% as beta carotene	*NP*
16,000 IU / 4,800 mcg RAE / 2 gummies (88% as beta carotene)	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*
32,000 IU / 9,600 mcg RAE / 2 softgels or 4 minis 88% as beta carotene	*NP*	*NP*	*NP*	32,000 IU / 9,600 mcg RAE / 2 softgels 88% as beta carotene	36,334 IU / 10,900 mcg RAE / 2 softgels 92% as beta carotene	36,334 IU / 10,900 mcg RAE / 2 softgels 92% as beta carotene	32,000 IU / 9,600 mcg RAE / 2 softgels 88% as beta carotene	36,334 IU / 10,900 mcg RAE / 2 softgels 92% as beta carotene
Vitamin D IU / mcg								
750 IU / 19 mcg / 0.5 ml	750 IU / 19 mcg / 1 ml	600 IU / 15 mcg / 1 ml	750 IU / 19 mcg / 1 ml	*NP*	*NP*	*NP*	*NP*	*NP*
1,500 IU / 38 mcg / 1 ml	1,500 IU / 38 mcg / 2 ml	1,200 IU / 30 mcg / 2 ml	1,500 IU / 38 mcg / 2 ml	*NP*	*NP*	*NP*	*NP*	*NP*
1,500 IU/ 37.5 mcg / 1 chewable	2,000 IU / 50 mcg / 1 chewable	1,200 IU / 30 mcg / 2 chewables	2,000 IU / 50 mcg / 1 chewable	3,000 IU / 75 mcg / 1 chewable	*NP*	*NP*	5,000 IU / 125 mcg / 1 chewable	*NP*
1,500 IU / 37.5 mcg / 2 gummies	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*
3,000 IU / 75 mcg / 2 softgels or 4 minis	*NP*	*NP*	*NP*	6,000 IU / 150 mcg / 2 softgels	6,000 IU / 150 mcg / 2 softgels	6,000 IU / 150 mcg / 2 softgels	10,000 IU / 250 mcg / 2 softgels	10,000 IU / 250 mcg / 2 softgels
Vitamin E IU / mg								
50 IU / 34 mg / 0.5 ml	50 IU / 34 mg / 1 ml[2]	50 IU / 34 mg / 1 ml[2]	50 IU / 34 mg / 1 ml[2]	*NP*	*NP*	*NP*	*NP*	*NP*
100 IU / 67mg/ 1 ml	100 IU / 67 mg / 2 ml[2]	100 IU / 67 mg / 2 ml[2]	100 IU / 67 mg / 2 ml[2]	*NP*	*NP*	*NP*	*NP*	*NP*
200 IU / 134 mg / 1 chewable	100 IU / 67 mg / 1 chewable[2]	100 IU / 67 mg / 2 chewables[2]	100 IU / 67 mg / 1 chewable[2]	200 IU / 134 mg / 1 chewable	*NP*	*NP*	200 IU / 134 mg / 1 chewable	*NP*
200IU / 134 mg / 2 gummies	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*
400 IU / 268 mg / 2 softgels or 4 minis	*NP*	*NP*	*NP*	400 IU / 268 mcg / 2 softgels	300 IU / 201 mg / 2 softgels[2]	300 IU / 201 mg / 2 softgels[2]	400 IU / 268 mg / 2 softgels	300 IU / 201 mg / 2 softgels
Vitamin K mcg								
500 / 0.5 ml	500 / 1 ml	400 / 1 ml	500 / 1 ml	*NP*	*NP*	*NP*	*NP*	*NP*
1,000 / 1 ml	1,000 / 2 ml	800 / 2 ml	1,000 / 2 ml	*NP*	*NP*	*NP*	*NP*	*NP*
1,000 / 1 chewable	1,000 / 1 chewable	700 / 2 chewables	1,000 / 1 chewable	1,000 / 1 chewable	*NP*	*NP*	1,000 / 1 chewable	*NP*
800 / 2 gummies	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*
1,600 / 2 softgels 2,000 / 4 minis	*NP*	*NP*	*NP*	1,600 / 2 softgels	2,000 / 2 softgels	2,000 / 2 softgels	1,600 / 2 softgels	2,000 / 2 softgels
Zinc mg								
5 / 0.5 ml	5 / 1 ml	5 / 1 ml	5 / 1 ml	*NP*	*NP*	*NP*	*NP*	*NP*
10 / 1 ml	10 / 2 ml	10 / 2 ml	10 / 2 ml	*NP*	*NP*	*NP*	*NP*	*NP*
15 / 1 chewable	10 / 1 chewables	10 / 2 chewables	10 / 1 chewables	15 / 1 chewable	*NP*	*NP*	15 / 1 chewable	*NP*
10 / 2 gummies	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*	*NP*
20 / 2 softgels or 4 minis	*NP*	*NP*	*NP*	20 / 2 softgels	20 / 2 softgels	20 / 2 softgels	20 / 2 softgels	20 / 2 softgels

WATER-SOLUBLE VITAMINS & ANTIOXIDANTS

CFF SPECIFIC VITAMINS				STEP 1			STEP 2	
MVW Complete Formulation® Drops, Chewables, Softgels / Softgel Minis	**GenADEK®** Drops, Chewables (Q3 2021)	**AquADEKs** Drops, Chewables	**DEKAs Plus** Drops, Chewables	**MVW Complete Formulation®** D3000 Chewables &Softgels	**GenADEK® STEP 1** Softgels	**DEKAs Plus** Softgels	**MVW Complete Formulation®** D5000 Chewables &Softgels	**GenADEK® STEP 2** Softgels
Thiamin B1 mg								
0.5 / 0.5 ml	0.6 / 1 ml	0.6 / 1 ml	0.6 / 1 ml	*NP*	*NP*	*NP*	*NP*	NP
1 / 1 ml	1.2 / 2 ml	1.2 / 2 ml	1.2 / 2 ml	*NP*	*NP*	*NP*	*NP*	NP
1.5 / 1 chewable	1.5 / 1 chewable	1.5 / 2 chewables	1.5 / 1 chewable	1.5 / chewable	*NP*	*NP*	1.5 / 1 chewable	NP
3 / 2 softgels or 4 minis	*NP*	*NP*	*NP*	3 / 2 softgels	3 / 2 softgels	3 / 2 softgels	3 / 2 softgels	3 / 2 softgels
Riboflavin B2 mg								
0.6 / 0.5 ml	0.6 / 1 ml	0.6 / 1 ml	0.6 / 1 ml	*NP*	*NP*	*NP*	*NP*	*NP*
1.2 / 1 ml	1.2 / 2 ml	1.2 / 2 ml	1.2 / 2 ml	*NP*	*NP*	*NP*	*NP*	*NP*
1.7 / 1 chewable	1.7 / 1 chewable	1.7 / 2 chewables	1.7 / 1 chewable	1.7 / 1 chewable	*NP*	*NP*	1.7 / 1 chewable	*NP*
3.4 / 2 softgels or 4 minis	*NP*	*NP*	NP	3.4 / softgels	3.4 / 2 softgels	3.4 / 2 softgels	3.4 / 2 softgels	3.4 / 2 softgels
mg Niacin Equivalents (NE)								
6 / 0.5 ml	6 / 1 ml	6 / 1 ml	6 / 1 ml	*NP*	*NP*	*NP*	*NP*	*NP*
12 / 1 ml	12 / 2 ml	12 / 2 ml	12 / 2 ml	*NP*	*NP*	*NP*	*NP*	*NP*
10 / 1 chewable	10 / 1 chewable	10 / 2 chewables	10 / 1 chewable	10 / 1 chewable	*NP*	*NP*	10 / 1 chewable	*NP*
40 / 2 softgels / 4 minis	*NP*	*NP*	*NP*	40 / 2 softgels	20 / 2 softgels	20 / 2 softgels	40 / 2 softgels	20/ 2 softgels
Pyridoxine B6 mg								
0.6 / 0.5 ml	0.6 / 1 ml	0.6 / 1 ml	0.6 / 1 ml	*NP*	*NP*	*NP*	*NP*	*NP*
1.2 / 1 ml	1.2 / 2 ml	1.2 / 2 ml	1/2 / 2 ml	*NP*	*NP*	*NP*	*NP*	*NP*
1.9 / 1 chewable	1.9 / 1 chewable	1.9 / 2 chewables	1.9 / 1 chewable	1.9 / 1 chewable	*NP*	*NP*	1.9 / 1 chewable	*NP*
3.8 / 2 softgels / 4 minis	*NP*	*NP*	*NP*	3.8 / 2 softgels	3.8 / 2 softgels	3.8 / 2 softgels	3.8 / 2 softgels	3.8 / 2 softgels
B12 mcg – Liquid Formulations Do Not Contain B12								
6 / 1 chewable	12 / 1 chewable	12 / 2 chewables	12 / 1 chewable	6 / 1 chewable	*NP*	*NP*	6 / 1 chewable	*NP*
12 / 2 softgels or 4 minis	*NP*	*NP*	*NP*	12 / 2 softgels	24 / 2 softgels	24 / 2 softgels	12 / 2 softgels	24 / 2 softgels
Biotin mcg								
15 / 0.5 ml	15 / 1 ml	15 / 1 ml	15 / 1 ml	*NP*	*NP*	*NP*	*NP*	*NP*
30 / 1 ml	30 / 2 ml	30 / 2 ml	30 / 2 ml	*NP*	*NP*	*NP*	*NP*	*NP*
100 / 1 chewable	100 / 1 chewable	100 / 2 chewables	100 / 1 chewable	100 / 1 chewable	*NP*	*NP*	100 / 1 chewable	*NP*
200 / 2 softgels or 4 minis	*NP*	*NP*	*NP*	200 / 2 softgels	200 / 2 softgels	200 / 2 softgels	200 / 2 softgels	200 / 2 softgels
Folic Acid mcg / mcg Dietary Folate Equivalents (DFE) – Liquid Formulations Do Not Contain Folic Acid								
200 mcg / 333 mcg DFE / 1 chewable	200 mcg / 333 mcg DFE / 1 chewable	200 mcg / 333 mcg DFE / 2 chewables	200 mcg / 333 mcg DFE / 1 chewable	200 mcg / 333 mcg DFE / 1 chewable	*NP*	*NP*	200 mcg / 333 mcg DFE / 1 chewable	*NP*
400 mcg / 667 mcg DFE / 2 softgels or 4 minis	*NP*	*NP*	*NP*	400 mcg / 667 mcg DFE / 2 softgels	400 mcg / 667 mcg DFE / 2 softgels	400 mcg / 667 mcg DFE / 2 softgels	400 mcg / 667 mcg DFE / 2 softgels	400 mcg / 667 mcg DFE / 2 softgels
Ascorbic Acid C mg								
45 / 0.5 ml	45 / 0.5 ml	45 / 1 ml	45 / 1 ml	*NP*	*NP*	*NP*	*NP*	*NP*
90 / 1 ml	90 / 1 ml	90 / 2 ml	90 / 2 ml	*NP*	*NP*	*NP*	*NP*	*NP*
100 / 1 chewable	70 / 1 chewable	70 / 2 chewables	70 / 1 chewable	100 / 1 chewable	*NP*	*NP*	100 / 1 chewable	*NP*
200 / 2 softgels or 4 minis	*NP*	*NP*	*NP*	200 / 2 softgels	150 / 2 softgels	150 / 2 softgels	200 / 2 softgels	150 / 2 softgels
Pantothenic Acid mg								
3 / 0.5 ml	3 / 1 ml	3 / 1 ml	3 / 1 ml	*NP*	*NP*	*NP*	*NP*	*NP*
6 / 1 ml	6 / 2 ml	6 / 2 ml	6 / 2 ml	*NP*	*NP*	*NP*	*NP*	*NP*
12 / 1 chewable	12 / 1 chewable	12 / 2 chewables	12 / 1 chewable	12 / 1 chewable	*NP*	*NP*	12 / 1 chewable	*NP*
24 / 2 softgels or 4 minis	*NP*	*NP*	*NP*	24 / 2 softgels	24 / 2 softgels	24 / 2 softgels	24 / 2 softgels	24 / 2 softgels
Selenium mcg								
-	10 / 1 ml	10 / 1 ml	10 / 1 ml	-	*NP*	*NP*	*NP*	*NP*
-	20 / 2 ml	20 / 2 ml	20 / 2 ml	-	*NP*	*NP*	*NP*	*NP*
-	75 / 1 chewable	75 / 2 chewables	75 / 1 chewable	-	*NP*	*NP*	-	*NP*
-	*NP*	*NP*	*NP*	-	150 / 2 softgels	150 / 2 softgels	-	150 / 2 softgels
CoQ10 mg								
-	2 / 1ml	2 / 1ml	2 / 1ml	-	*NP*	*NP*	*NP*	*NP*
-	4 / 2 ml	4 / 2 ml	4 / 2 ml	-	*NP*	*NP*	*NP*	*NP*
-	10 / 1 chewable	10 / 2 chewables	10 / 1 chewable	-	*NP*	*NP*	-	*NP*
-	*NP*	*NP*	*NP*	-	20 / 2 softgels	20 / 2 softgels	-	20 / 2 softgels

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5. DEKAs are manufactured by Callion Pharma, MVW Complete Formulation is a registered trademark of MVW Nutritionals, Inc., GenADEK is a registered trademark of Generix Development Group, Inc.
6. MVW ADEK Gummies contain fat-soluble vitamins and zinc

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Chapter 17

NUTRITION INTERVENTIONS FOR CONGENITAL HEART DISEASE

Deonna Monnahan, MS, RDN, CD, CNSC

Congenital heart disease (CHD) refers to cardiovascular defects that are present at birth. CHD is the most common type of birth defect (1). From 2010-2017, the worldwide incidence was 9.4 out of 1000 births (1). Although some genetic abnormalities have increased risk of heart disease, most often the cause of CHD is unknown. There are many different kinds of CHD, and they can vary in severity from mild to severe (see Table 17-1) (2). The most common type is ventricular septal defect (VSD) (2). CHD lesions can be categorized as cyanotic (not enough oxygen in the blood, which can result in a bluish color to skin and mucous membranes) or acyanotic (oxygen content of blood is not normally affected), with acyanotic being more common (3). The treatment for CHD can vary greatly from no intervention needed to medications, cardiac catheterizations, surgery, or heart transplant.

Table 17-1: Severity of CHD lesions (2):

Severe	Moderate	Mild
D-transposition of the great arteries	Mild or moderate aortic stenosis or aortic incompetence	Small ventricular septal defect
Tetralogy of Fallot	Moderate pulmonic stenosis or incompetence	Small patent ductus arteriosus
Hypoplastic right heart	Noncritical Coarc	Mild pulmonic stenosis
Hypoplastic left heart	Large atrial septal defect	Bicuspid aortic valve without aortic stenosis or aortic incompetence
Single ventricle	Complex forms of ventricular septal defect	Small or spontaneously closed atrial septal defect
Double outlet left ventricle		
Truncus arteriosus		
Total anomalous pulmonary venous connection		
Critical pulmonic stenosis		
Atrioventricular septal defect		
Large ventricular septal defect		
Large patent ductus arteriosus		
Critical or severe aortic stenosis		
Severe pulmonic stenosis		
Critical Coarc		

CHD and malnutrition

Children with CHD are at risk for malnutrition, which can negatively impact surgical outcomes and increase risk for morbidity and mortality (4). Infants with CHD are also at risk for neurodevelopmental delay (5). Children with CHD may have simultaneously slow growth velocity for weight, length, and head circumference during infancy and toddler years (6). Adequate nutrition is crucial to support growth and typical development, optimize surgical outcomes, and reduce the risk of mortality (7). Table 17-2 lists the reasons CHD puts children at risk for malnutrition.

Table 17-2: Contributors to Poor Growth in Infants and Children with CHD (5, 6, 8)

Increased metabolic demands from heart disease	Tachycardia
Anorexia	Frequent periods of inadequate intake due to medical interventions
Oral feeding difficulties	Nutrient losses from protein-losing enteropathy or medication interactions
Vomiting	Neurologic injury
Gastroesophageal reflux disease (GERD)	Genetic abnormalities
Early satiety	
Tachypnea	

CHD and feeding difficulties

Infants and children with CHD may require the support of a feeding tube to provide nutrition support due to difficulties feeding by mouth. The main contributors to feeding difficulties are listed in Table 17-3.

Table 17-3: Contributors to Feeding Difficulties in Infants and Children with CHD (7)

Tachypnea or increased work of breathing	Medication side effects (nausea, vomiting, constipation, drowsiness)
Hypoxia	Neurological injury
Respiratory support	Genetic syndromes
Fatigue and inadequate endurance for feeding	Damage to vocal cords
GERD	Dysphagia
Medical interventions delaying opportunities for oral feeding during critical period of developing oral feeding skills during infancy	Oral aversion

Signs of feeding difficulties and possible aspiration

If an infant or child is showing any of these signs while feeding, ask the cardiologist or pediatrician for a referral to an occupational therapist/physical therapist/speech therapist that specializes in feeding.

- Poor coordination of sucking, swallowing, and breathing in infant
- Worsening of oxygen desaturations
- Increased heart rate
- Increased work of breathing
- Coughing
- Choking
- Gagging
- Retching
- Red/watery eyes
- Fluids spilling out of mouth
- Feeding refusal

Nutrition goals for infants and children with CHD

- Adequate nutrition intake to support normal and consistent weight gain and growth, or catch-up weight gain if needed. Sometimes surgeries are delayed until the infant or child reaches a certain weight.
- Normal electrolyte, vitamin and mineral levels.
- Support age-appropriate feeding behaviors if infant/child is safely able to participate.
- Limit feeding intolerance as much as possible.

Estimated needs

Severe	Moderate	Mild
Calories	110-140 kcal/kg/day	BMR x 1.5-1.8, but may need up to x2.2 with severe heart failure/ malnutrition
Protein	1.8-3.5 grams/kg/day	1-2.5 grams/kg/day, may need more with protein losing enteropathy
Fluid*	At least 100 mL/kg/day. Cardiologist may want to restrict to <145 mL/kg/day for infant that is tube fed and on diuretics. Infants fully orally feeding should not be fluid restricted and instead diuretics dosing may be adjusted by cardiologist.	Cardiologist may prescribe fluid restriction depending on kind of heart disease.

Note that with mild cardiac lesions or after recovery from surgical repair, calorie and protein needs may be similar to typically developing, healthy infants and children.

*Fluid retention and volume overload to the lungs or other organs can occur with CHD and may require fluid restriction and diuretics.

Sodium

Some children with fluid retention and edema may need to restrict their sodium intake. Occasionally an infant or child may be prescribed sodium supplements due to hyponatremia while on diuretics. Ask the child's cardiologist if you are unsure about the child's sodium needs.

Vitamin D

Children with CHD are likely to have lower serum vitamin D levels compared to typically developing children (9), and in particular those who underwent cardiopulmonary bypass are at risk for vitamin D deficiency (10). Additional vitamin D supplementation is necessary for those with diagnosed deficiency, along with a plan to recheck a vitamin D level to monitor for repletion.

Iron

Some children with CHD will be at risk for iron deficiency. Iron deficiency is common for children with heart failure, especially those with more advanced heart failure, and is associated with poor clinical outcomes (11). These children may have iron deficiency without anemia, and therefore it is important to check an iron profile (iron level, total iron binding capacity, iron saturation and ferritin). Typically the cardiologist will determine the dose of iron supplementation needed and plan for monitoring.

Strategies to increase calorie and protein intake for infants

Infants with CHD often require breast/chest milk fortification or increasing caloric concentration of formulas. Start with 22-24 calories/ounce and increase as needed and as tolerated by 2-3 calories/ounce at a time to a maximum of 30 calories/ounce. Fortification and formula recipes for parents and caregivers can be found on the Seattle Children's Hospital website: https://www.seattlechildrens.org/clinics/nutrition/patient-family-resources/

For infants who are fully breast/chestfeeding but need to increase their caloric intake, the number of episodes of daily breast/chestfeeding will need to be decreased in order to allow for opportunities to offer bottles of fortified breast/chest milk. Consider starting with 3-4 bottles a day of fortified breast/chest milk. These bottles may require increasing to a higher level of fortification (26-30 calories/ounce) to support weight gain while continuing breast/chestfeeding at the other feedings.

When choosing a formula, first choice should be a standard cow's milk protein-based formula unless the parent or caregiver has a strong preference for otherwise. If that is not well tolerated then consider a partially hydrolyzed formula, and then if needed an elemental formula. There are few indications for a soy formula, but a soy formula could be considered for a full-term infant who is fully orally feeding by mouth, cannot have cow's milk based formula and refuses partially

hydrolyzed/elemental formulas. Keep in mind there can be many reasons for feeding intolerance other than formulas for those with CHD, as previously mentioned.

Since infants with CHD often have high calorie needs, when their calorie needs are met with breast/chest milk fortification or concentrated formulas then usually their protein needs are met as well, and a protein supplement is rarely required.

When fortifying breast/chest milk or concentrating formulas, protein-free calorie additives such as Duocal or an oil additive are not needed unless the infant is requiring such a high amount of formula that they are receiving more than 4 g/kg/day protein. An exception to this would be if the infant has another diagnosis that requires limiting protein, such as certain types of kidney failure.

Strategies to increase calories and protein intake for children and teens

Encourage high calorie and high protein foods at each meal and snack, and calorie containing beverages. Focus on adding fats to meals and snacks since many children have early satiety and are not able to eat larger volumes of food.

Those with nausea or early satiety will benefit from small, more frequent meals and snacks.

Some children and teens may benefit from high calorie, commercial oral supplements, especially if homemade shakes and smoothies are not accepted or a feasible option for home.

Overweight and obesity

Recent research shows children with CHD have similar rates of overweight and obesity as the general population (12). A major contributor to overweight and obesity in children with CHD is physical inactivity, which may be due to real and/or perceived limitations. Some types of heart disease may warrant medically prescribed physical activity restrictions, while physical activity may also unnecessarily be limited due to child/parent/caregiver concerns about safety or misunderstanding the medical instructions (13). Another possible contributor is the impact of the focus on weight gain and nutrition interventions in infancy leading to overweight and obesity later in life (12).

Assessing growth

Use standard pediatric growth charts that apply to the child's age, while also incorporating syndrome specific growth charts when applicable, such as for Trisomy 21, Trisomy 18, Noonan syndrome and 22q11 deletion syndrome (see Chapter 2).

Serial measurements of mid-upper arm circumference (MUAC) over time can be a better indication of nutrition status for infants and children with edema that have unreliable weight changes (14). MUAC growth charts are available to determine z-score and presence of malnutrition.

Keep in mind how fluid shifts can affect weight changes. For some children, periods of rapid weight gain may reflect fluid retention and edema, and a brief period of weight loss can occur after diuretics are started or dosing is increased significantly.

Trouble shooting feeding intolerance

Feeding intolerance can occur with CHD due to poor gut perfusion (7).

Reflux	Ask the pediatrician or cardiologist if proton-pump inhibitor or histamine-2 blocker medications are appropriate. These medications do not stop reflux but will help alleviate pain. Unchecked pain from reflux can lead to oral aversion. For infants, a MD or OT/PT may recommend a wedge to help keep infant upright.
Vomiting	If infant is on a high amount of breast/chest milk fortification or formula concentration, consider a trial of decreasing density to assess tolerance. Possible intolerance to type of formula. If child is tube fed, consider overfeeding as possible cause, especially if child is still gaining weight despite vomiting. If child is tube fed, consider slowing down the rate of bolus feeds to be given over 1 hour or change to continuous feeds. Consider constipation as possible contributor. Consider medication side effects.
Constipation	Consider introducing small amounts of pear or prune juice to assist with bowel movement or asking pediatrician or cardiologist about starting bowel regimen medications. For >1 year old, assess fiber and fluid intake.
Diarrhea	Consider medication side effects as possible cause. Possible intolerance to type of formula.

Protein losing enteropathy

Protein losing enteropathy (PLE) as a result of CHD or the Fontan procedure is usually due to increased venous pressure causing an excessive loss of proteins from the lymph system into the GI tract (15). Some protein will be reabsorbed into the portal circulation, however PLE occurs when the amount of protein leaked into the gut exceeds the amount that can be reabsorbed (16). PLE diagnosis can be confirmed by high levels of alpha-1-antitrypsin in the stool (15). Development of PLE after Fontan procedure significantly increases the risk of mortality (15).

Children with PLE may have (15, 16):

- Hypoalbuminemia
- Edema
- Ascites
- Pleural or pericardial effusions
- Malnutrition
- Growth failure
- Diarrhea
- Nausea
- Vomiting
- Malabsorption, including of fat-soluble vitamins
- Electrolyte abnormalities

Treatment of PLE

Those with PLE should eat a high protein diet and may require the help of protein supplementation. Goal protein intake is typically ≥2 grams/kg/day (16). Serum levels of fat soluble vitamins should be monitored and vitamin supplementation should be given when deficiencies are found.

A low-fat diet (<25% of calories from fat) may be recommended with the goal of decreasing the lymphatic flow and pressure and thereby possibly limiting protein losses (16). With low-fat diets typically adding medium-chain triglyceride (MCT) oil is recommended since MCT is not absorbed into the lymph system and instead goes directly to the portal venous circulation (15). The purpose of MCT oil is to add more fat to the child's diet (it will not help to raise albumin levels), but it can be challenging to incorporate due to the cost, unpalatable taste and GI upset large amounts can cause. In this author's experience if a low-fat diet has resulted in the child losing weight or an overall poor nutrition intake then a trial of reintroducing fat (with approval by the cardiologist) is warranted to see if the change has any impact on albumin levels or other symptoms of PLE.

Reducing sodium intake is commonly needed for those with edema.

Note that diet changes alone rarely result in any improvement of PLE (16), and PLE may not resolve until the underlying cause is treated with medications, procedures and/or surgery, which for some will mean heart transplant.

Post-operative chylothorax

Chylothorax is a rare but serious condition that can happen after cardiac surgery, due to trauma to the thoracic duct or lymphatic vessels during surgery or increased venous pressure (17). Chylothorax results in chyle leaking from the lymphatic system into the pleural cavity (17). The chylous fluid typically needs to be removed by chest tube after surgery, during which important nutrients are lost, including fat, glucose, protein, fat soluble vitamins, electrolytes and minerals (17). Chyle is rich in fat since during digestion long-chain triglycerides are formed into chylomicrons, which enter the lymphatic system via the lymph vessels of the small intestine (18).

Treatment of chylothorax

Nutrition interventions focus on a low-fat diet in order to decrease the amount of chyle that travels through the lymph system to allow healing of the leak (18). The low-fat diet is typically <25% calories from fat, but further reduction may be needed depending on the child or direction of cardiologist. In severe post-op cases a period of total parenteral nutrition as sole source nutrition is needed. The low fat-diet is typically continued after the leak is resolved, for a period of time defined by the cardiologist, but usually at least 4 weeks.

Similar to nutrition interventions for PLE, MCT oil supplementation is usually recommended since it does not travel through the lymph system. Infants with chylothorax will require a formula low in long-chain triglycerides and high in medium-chain triglycerides, such as Enfaport, Lipistart or Monogen. The MCT content of these formulas is crucial for the growth of infants, whereas older children on a low-fat diet could go without the addition of MCT oil if not well accepted or tolerated.

Infants fed breast/chest milk could receive skimmed breast/chest milk fortified with these specialty formulas during treatment for chylothorax, which requires the parent or caregiver to receive careful instruction in the hospital on how to properly skim the breast/chest milk at home. These formulas are unpalatable and therefore some infants may refuse to take them by mouth and may require the temporary assistance of a feeding tube.

Older children who are tube fed will also require these specialty formulas or a blended tube feeding recipe that is low in fat. Essential fatty acid requirements can be met if at least 2-4% of total caloric intake is from linoleic acid (an essential n-6 polyunsaturated fatty acid) (18). MCT oil does not contain essential fatty acids.

If vitamin deficiencies were discovered while inpatient then serum levels will need to be rechecked outpatient as well until repleted.

Table 17-4: Common medication interactions

Common medications	Possible Nutrition-related Impacts	Nutrition Interventions
Diuretics: (19) Loop diuretics: Furosemide Bumetanide Thiazide diuretics: Hydrochlorothiazide Chlorothiazide Metolazone Potassium Sparing: Spironolactone	All of these diuretics impact electrolyte levels and acid-base balance. Loop diuretics: cause losses of sodium, potassium, chloride, magnesium, and calcium. High losses of calcium through urine can for infants lead to nephrocalcinosis, nephrolithiasis or decreased bone density. Thiazide diuretics: cause losses of sodium, potassium, chloride and magnesium. Increases calcium reabsorption, which may help with bone density, but could also increase risk for hypercalcemia. Potassium Sparing: can increase risk for hyperkalemia.	Ensure infant/child is receiving at least DRIs for calcium and vitamin D intake. Monitor serum vitamin D level given increased risk for bone demineralization. Cardiologist will monitor electrolyte and mineral levels and prescribe supplements as needed.
Antibiotics	May cause diarrhea.	Consider adding probiotics from foods or a supplement, such as lactobacillus.
Steroids: (20, 21) Prednisone Prednisolone Methylprednisolone Hydrocortisone Dexamethasone	Osteopenia Growth restriction Muscle loss Swollen or puffy face Fluid retention/edema Increased hunger and weight gain Stomach discomfort At risk for gastric ulcer Hyperglycemia Hyperlipidemia Slowed growth Delayed puberty Mood swings Acne Increased risk of infection Poor wound healing Hypertension	Ensure adequate calcium and vitamin D intake to protect bones. Seattle Children's Hospital has a resource on recommended intake: https://www.seattlechildrens.org/pdf/PE615.pdf Encourage high protein foods and physical activity to minimize muscle loss. Sodium restriction may be needed for those with edema. Diet modification needed for those with hyperglycemia and hyperlipidemia.
Electrolyte supplementation: Sodium, potassium or chloride supplements	Vomiting	Notify cardiologist if vomiting is significant because dosing of supplements or diuretics may need to be adjusted. Suggest foods/beverages high in these electrolytes to hopefully reduce amount of supplementation needed (does not apply to infants).

continued...

Common medications (continued)	Possible Nutrition-related Impacts	Nutrition Interventions
Anticoagulants: Warfarin Heparin	At risk for bleeding or clotting with sudden changes to vitamin K intake. Bone loss/decreased bone density with long-term use (22).	With warfarin it is necessary to follow a vitamin K consistent diet. Each medical center has their own approach to this, for which the child's anticoagulation pharmacist will be an important resource. https://www.seattlechildrens.org/globalassets/documents/for-patients-and-families/pfe/pe3314.pdf Goal of receiving at least DRI for calcium and vitamin D intake to protect bones. With long-term use consider check vitamin D level.
Proton pump inhibitor and histamine-2 blocker (22)	May reduce calcium absorption and therefore have negative impact on bone health.	Goal of receiving at least DRI for calcium and vitamin D intake to protect bones.
Immune suppression drugs for after heart transplant: Mycophenolate Mofetil (MMF) Cyclosporine Tacrolimus	MMF: nausea, vomiting, and diarrhea. Cyclosporine: Hypertension, hyperlipidemia, nausea, vomiting, osteoporosis. Grapefruit must be avoided due to drug-nutrient interaction. Causes losses of magnesium. Tacrolimus: hypertension and hyperlipidemia. Grapefruit must be avoided due to drug-nutrient interaction. Causes losses of magnesium.	Should follow immune suppressed diet. See these resources from Seattle Children's Hospital or USDA for more information: https://www.seattlechildrens.org/pdf/PE2372.pdf https://www.fsis.usda.gov/sites/default/files/media_file/2021-04/at-risk-booklet.pdf Avoid all sources of grapefruit with cyclosporine and tacrolimus. Goal of receiving at least DRI for calcium and vitamin D intake to protect bones. Monitor serum vitamin D level given increased risk for bone demineralization. Magnesium supplementation may be needed with cyclosporine and tacrolimus. Encourage foods rich in magnesium, but often supplementation is still needed. Diet modifications may be needed with hypertension and hyperlipidemia.
Mineral supplementation: Magnesium Calcium Iron	Magnesium supplements can cause diarrhea. Iron supplements can cause constipation.	Suggest foods high in magnesium and calcium to hopefully reduce amount of supplementation needed. See constipation section.

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Chapter 18

NUTRITION INTERVENTIONS FOR INTESTINAL FAILURE AND SHORT BOWEL SYNDROME

Nila Williamson MPH, RDN, CNSC

Definition of Intestinal Failure and Short Bowel Syndrome

Intestinal failure is defined as "the reduction of functional gut mass below the minimal amount necessary for digestion and absorption adequate to satisfy the nutrient and fluid requirements for maintenance in adults or growth in children (1). Intestinal failure includes individuals with surgical short bowel syndrome, dysmotility (e.g. intestinal pseudoobstruction), and those born with rare congenital enterocyte disorders (e.g. microvillus inclusion disease, etc.). In children, the most common cause of intestinal failure is short bowel syndrome. Short bowel syndrome occurs when a significant length of the bowel is surgically removed or resected, most commonly during the newborn period, to treat conditions such as necrotizing enterocolitis, malrotation, volvulus, gastroschisis, atresia, or Hirschsprung's disease. Short bowel syndrome can also occur in older children and adults following trauma to the bowel such as after an auto accident, fall, or severe non-accidental trauma (2).

Anatomy of the Bowel

Small bowel (small intestine) length is one of many factors that help to predict weaning from total parenteral nutrition support (TPN) following bowel resection (3). Several studies have sought to measure typical small bowel length using a variety of methods. A recent study measured small bowel length in live preterm infants and children up to the age of 5 years during a laparotomy procedure and found that small bowel length doubles during the last trimester of pregnancy. Preterm infants at 24-27 weeks had an average small bowel length of 100 cm, compared to term infants of 40 weeks having 150-200 cm. By the age of 5 years children had an average of 424 cm. (4).

The small intestine consists of the duodenum, jejunum, and ileum. (See Figure 18-1). The portion of bowel resected will influence nutritional concerns. The duodenum is the first portion of the small bowel from the pyloric valve to the ligament of Treitz. The Jejunum consists of the proximal two-fifths of the small bowel past the ligament of Treitz, and the ileum is the distal three-fifths of

the small bowel to the ileocecal valve. The majority of carbohydrate and protein absorption takes place in the duodenum and jejunum. Fats and fat-soluble vitamins are absorbed in the ileum. Bile salts are excreted from the liver into the duodenum and are required for the absorption of long chain fatty acids and fat-soluble vitamins in the ileum. Vitamin B12 binds to intrinsic factor (produced in the stomach) and is absorbed in the terminal ileum. Fluids and electrolytes are predominantly absorbed in the ileum and in the colon. When the duodenum and/or jejunum are resected, the ileum can largely adapt to perform their absorptive functions. The duodenum and jejunum, however, cannot adapt to perform the functions of the ileum.

The ileocecal valve is the main barrier between the small and large intestine. It helps regulate the release of fluid into the colon and prevents reflux of colonic bacteria into the small bowel. The ileocecal valve is an important factor in outcomes for children with short bowel syndrome. Resection of the ileocecal valve results in decreased fluid and nutrient absorption, and increased bacterial overgrowth in the small bowel. The presence of the ileocecal valve is positive predictor of weaning off of TPN (5).

The colon, or large intestine, is located between the ileocecal valve and the rectum. Its primary function is the absorption of fluid, electrolytes, and storage of stool. Complications of its resection include dehydration, electrolyte abnormalities, and reduced absorption of bile salts for fat absorption.

Stages of Short Bowel Syndrome

Short bowel syndrome is characterized by three phases. The first is the early post-operative phase. During this time TPN is the sole nutrition source. High fluid and electrolyte losses from ostomies or diarrhea are common, and therefore fluid resuscitation and replacement are needed to prevent dehydration and electrolyte abnormalities.

The second stage is adaptation. Adaptation is the term used to describe the process by which the remaining bowel grows and compensates to take on the nutrition and fluid absorption of the portion of bowel that was lost. This stage is characterized by cellular proliferation and angiogenesis (development of new blood vessels) and occurs over a period of months to years, particularly for infants and children. During adaptation, enteral feeds should be initiated either with frequent small volume oral feedings or continuous drip tube feeding. Feeding volumes are slowly advanced so as not to overwhelm the remaining bowel's absorptive capacity. Adaptation and structural changes to the bowel such as brush border enzyme activity are stimulated and maximized by the use of enteral nutrition. During this phase, TPN is weaned as the bowel adapts and absorption of enteral intake increases to maintain appropriate growth (3).

The third and final phase of short bowel syndrome is equilibrium. Once this stage is reached there is no further bowel adaptation. It is the goal that before this is reached the child has been

able to achieve enteral autonomy, or independence from TPN. For those patients that cannot achieve enteral autonomy, the equilibrium phase is characterized by adjusting TPN to maintain appropriate growth, monitor for nutrient deficiencies and liver function, and manage to minimize or avoid development of liver disease.
Factors that will determine how well an infant with short bowel syndrome will fare and how much enteral nutrition they will go on to tolerate include:

- Age at the time of intestinal surgery with infants having the greatest future growth potential of the remaining bowel
- Amount of bowel resected and site of resection (duodenum, jejunum, ileum, ileocecal valve, colon)
- Function (absorptive and motility) of the remaining bowel
- Adaptive capacity of the remaining bowel
- Injury to the bowel in the forms of bacterial overgrowth, infections, ischemia, etc.
- Occurrence of complications from long-term TPN (e.g. liver disease, recurrent line infections, loss of central access) (6)

Nutritional Support in Short Bowel Syndrome

Immediately after a bowel surgery which results in short bowel syndrome, TPN is required until bowel function returns (bowel sounds are detected and stool is produced). Depending on the severity of short bowel syndrome, full enteral/oral nutrition may be achieved in a matter of weeks, months, or may never be achieved. A need for parenteral nutrition longer than 90 days defines one as having intestinal failure (7). It is important that a patient be given as much enteral/oral nutrition as tolerated to facilitate bowel adaptation. Enteral intake is also important for protecting against liver disease (6).

Introduction of oral feedings are important for development and prevention of oral feeding aversion. Small volume oral feedings of breast milk or formula should be introduced as soon as an infant is stable after surgery, and then increased gradually as tolerated. If oral skills are not present, trophic volumes of continuous tubefeeding with breastmilk or formula should be initiated and used to supplement what the child cannot take by mouth. Evidence is lacking on the optimal type of formula feeding, mode of feeding (continuous versus bolus), and route of delivery (oral versus nasogastric versus gastrostomy versus post-pyloric) for children with intestinal failure and short bowel syndrome (3). Guidelines and recommendations in this area are therefore based more on clinical practice and experience, than published evidence.

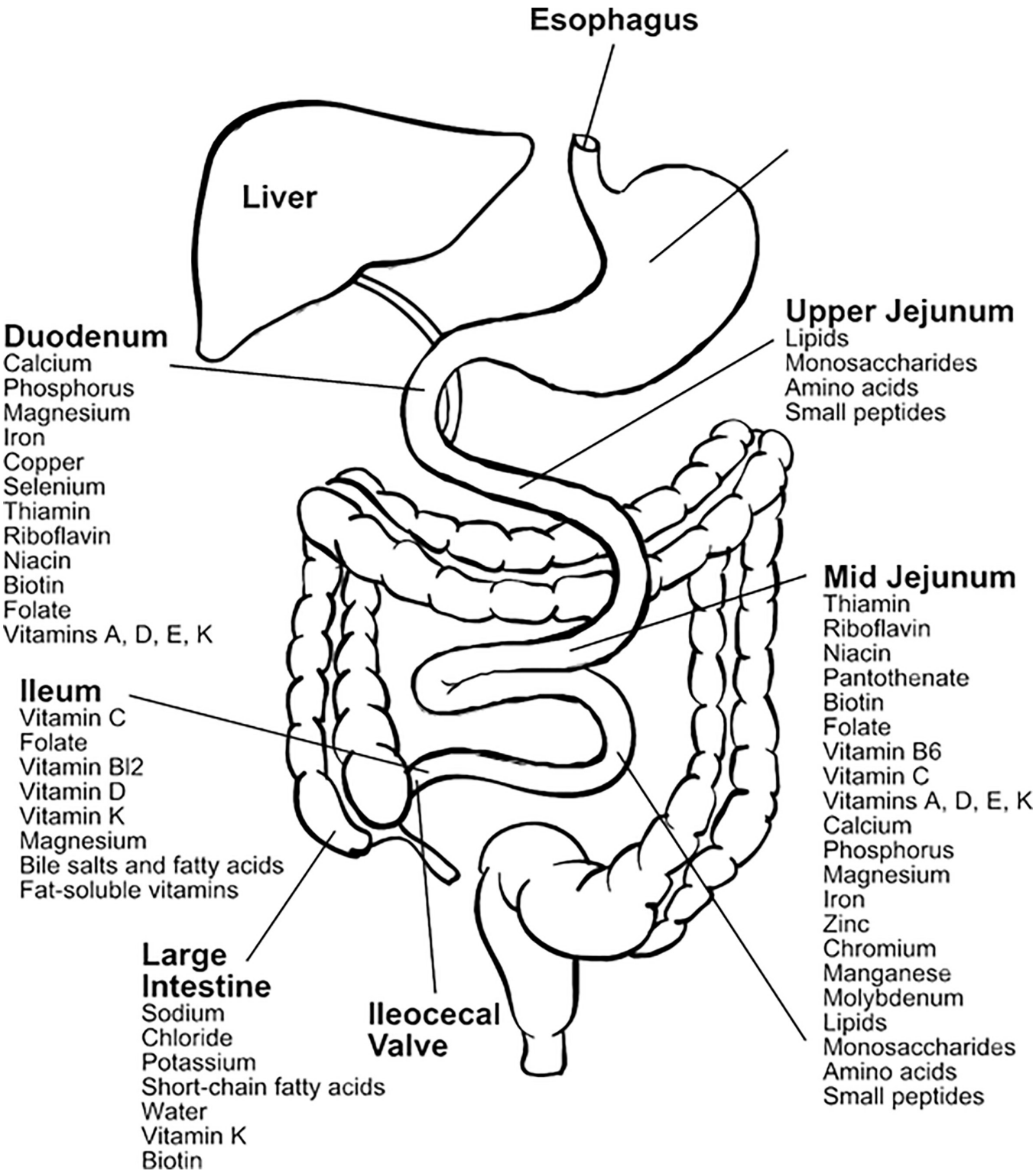
Breastmilk is the ideal first food for infants with short bowel syndrome because of its human milk oligosaccharide (HMO) content that serves as prebiotics and immunomodulators capable of stimulating gut adaptation and preventing infection (8). Extra breastfeeding support and education should be provided by the medical team to the mothers of these infants because of its health benefits for the infant. When breastmilk is not available formula should be substituted.

Historically amino-acid-based formulas have been favored for children with short bowel syndrome due to their hypoallergenicity and inclusion of medium chain triglycerides (2). It is thought that the impaired gut function of children with intestinal failure may increase their risk of allergic gastrointestinal diseases and medium chain triglycerides can be absorbed anywhere in the small intestinal as they do not require bile salts for absorption. However more recently some intestinal rehabilitation centers are transitioning infants to standard intact protein formulas once breastmilk is no longer available and monitoring the child's tolerance (9). A benefit of using intact protein formulas is the maintenance of oral intake. Amino-acid based formulas are well known for not being very palatable and oral acceptance of these formulas can be difficult for many infants. Additionally, animal studies suggest intact macronutrients may promote better intestinal adaptation, but data from human studies is limited (10).

Solid foods should be introduced when developmentally appropriate (2). All fruits and simple sugars should be strictly avoided because of carbohydrate malabsorption and their ability to loosen stools, further worsening diarrhea. With these exceptions, a variety of vegetables, whole grain cereals, and infant proteins should be introduced. Often the fiber and protein in these foods will help slow transit time and thicken stool output. Textures should be advanced to table foods as child is capable. It is important to involve feeding therapists early if there is a concern for a delay in feeding skills (6).

Children with short bowel syndrome receiving enteral/oral intake have large volumes of loose stools. Increases in intake of breastmilk or formula by mouth or by tube should be made very slowly and in a controlled manner; with increases only once per week or every 3-4 days when the child is very young. Changes in stool output (consistency and volume) should be monitored with increases in intake. Typically an upper limit of eight to ten watery stools per day, any skin breakdown or diaper rash, or ostomy output greater than ~40 mL/kg/day is used as a holding point for feeding advancement. Weight gain and hydration should also be closely monitored. Once stool output improves, advancement of feeds can resume. Slow and steady feeding advancement that is tolerated without excessive stool output is better than overwhelming the bowel's absorptive capacity and having to stop and start feeding routines. The stability of slow and steady feeding advancement methods also leads to improved safety for making TPN reductions. As larger volumes of oral/enteral intake are tolerated and weight gain is exceeding expected rates for age, TPN should be reduced. It is important not to over nourish a child with TPN as this is a risk factor for liver disease (11).

Figure 18-1: Intestinal Tract - Sites of Nutrient Absorption



Complications in Short Bowel Syndrome

Micronutrient Deficiencies

Children with intestinal failure transitioning from parenteral nutrition to oral/enteral nutrition experience a high prevalence of micronutrient deficiencies (12). The site of bowel resection will help identify nutrients of concern. Duodenal resection will increase risk for iron deficiency. Ileal resection can result in fat and fat-soluble vitamin malabsorption as well as B12 deficiency. It is frequently necessary to give high doses of fat-soluble vitamins in an absorbable water-soluble form. Following ileal resection children will require lifelong Vitamin B12 supplementation in the form of a sublingual tablet, nasal spray, or intramuscular injection. Tablets designed to be swallowed will not be absorbed due to the absence of ileum. It can take several months to several years for a vitamin B12 deficiency to develop; therefore, long-term, regular monitoring of B12 status is necessary (13). Common nutrient deficiencies due to malabsorption from short bowel syndrome also include calcium, Vitamin D, Vitamin E, iron, magnesium, and zinc (2). Essential fatty acid deficiency is also a risk (14). These nutrients need to be monitored routinely for the patient's entire life, but especially and more frequently in the months just after parenteral nutrition is discontinued (6).

More recently, shortages of intravenous micronutrient and electrolyte preparations are occurring. During these shortages, patients still dependent on TPN and unable to tolerate enteral supplementation are at risk for deficiencies and subsequent adverse outcomes (10).

Bacterial Overgrowth

Children with short bowel syndrome often have poor intestinal motility and dilated segments of the small intestine where contents can pool and ferment. This, plus absence of the ileocecal valve and acid suppression, contribute to the development of bacterial overgrowth. Bacterial overgrowth is present when the bacteria in the small bowel exceed normal levels and cause unpleasant symptoms. Bacterial overgrowth results in malabsorption by causing inflammation of the bowel wall. Symptoms vary between individuals but can include weight loss, very foul-smelling stools and flatus, bloating/distention, diarrhea, vomiting, and in severe cases, bleeding intestinal ulcers, and accumulation of D-lactic acid in the blood. Bacterial overgrowth may also increase intestinal permeability, resulting in an increased risk for development of central line infections. One study looking at this risk found that infants with short bowel syndrome and bacterial overgrowth had 7 times the rate of central line blood stream infections than those infants with short bowel syndrome and no bacterial overgrowth (15). Bacterial overgrowth can be diagnosed by breath hydrogen test, aspiration and culture of small bowel contents, or by blood test for D-lactic acid. However due to the difficulty in administering these tests it is commonly diagnosed based on clinical symptoms. Bacterial overgrowth is treated with oral antibiotics. For some patients cyclic antibiotic schedules or continuous antibiotics are necessary. In these cases, antibiotics may be rotated periodically to avoid overgrowth of resistant bacteria (5, 6).

Central Line Infections and Vascular Access

Central line care is a critical component of the nutrition management of children with intestinal failure because they require central venous access for long-term TPN. Extensive education on aseptic technique and care of the line to prevent breaking is a requirement for caregivers before they begin TPN at home. Central line bloodstream infections are a major contributor to morbidity and mortality of the pediatric intestinal failure population. These infections, when occurring frequently, are associated with a higher risk for parenteral nutrition associated liver disease. Incidence of central line bloodstream infections in this population has been reported as high as 80%. Ethanol lock therapy is proven for reducing infections. A 70% ethanol solution is instilled into the central line and left there during time the line is not being used for TPN administration. In order to receive ethanol locks the child needs to be old enough or tolerating sufficient oral/enteral feedings to have a break in time each day from TPN (5, 6, 16).

Liver Disease

Children with intestinal failure who depend on long term TPN are susceptible to the development of parenteral nutrition associated liver disease (PNALD). Two-thirds of patients with intestinal failure will develop PNALD, and historically 25% advanced to end-stage liver disease. Despite the favorable long-term survival rate of 70-90%, the goal of medical management for children with intestinal failure is to promote intestinal adaptation and intestinal function while preventing the development of irreversible liver damage (16). Risk factors for development of PNALD include premature birth, fasting resulting in impaired bile flow, early or recurrent catheter-related sepsis, and the macronutrient composition of TPN (particularly lipid). Strategies protective of PNALD include early introduction of enteral feeding, reduction of TPN to prevent overfeeding, lipid minimization, cycling of TPN to allow several hours off TPN each day, and prevention of central line bloodstream infections by using strict aseptic care techniques.

Although excesses of any macronutrient (glucose, protein, and lipid) contribute to stress on the liver, lipid is the single intravenous nutrient that most contributes to the development of PNALD. The strategies of lipid minimization to 1 g/kg/day and replacement of soy-based lipids with fish oil-based lipids or mixed-lipid emulsions has significantly reduced the prevalence of PNALD (3). Mixed-lipid emulsions contain a blend of soy, MCT, olive, and fish oils which offers a more balanced fatty acid profile. This may provide a solution to the current challenges in meeting energy needs with lipid minimization, and challenges with essential fatty acid deficiencies when having to choose between predominantly omega-6 soy lipid or omega-3 fish oil lipids. Studies are ongoing (17).

Conclusion

Outcomes and survival rates for children with intestinal failure have improved dramatically over the past half century. Survival has risen from 54% prior to 1972 to 94% at this time. (3). It remains important to stress however, that children with intestinal failure and short bowel syndrome require lifelong nutritional care. Children dependent on TPN as well as those that have transitioned to being enterally autonomous need close monitoring. Any time they have an illness resulting in increased stool or ostomy output, they are at high risk for dehydration with fluid and electrolyte imbalances. They need long term, regular nutrition monitoring to prevent problems associated with macro-and micronutrient malabsorption, which can result in poor weight gain, growth, and nutrient deficiencies. Care by a multidisciplinary intestinal rehabilitation team, that considers a child's nutritional, medical, and surgical needs, is associated with increased survival, improved outcomes, and decreased need for intestinal transplantation (2, 3, 5, 10).

The remainder of this chapter presents guidelines for nutrition assessment, intervention, and evaluation/outcome for children with short bowel syndrome.

Table 18-1: Nutrition Interventions for Short Bowel Syndrome

Assessment	Intervention	Evaluation/Outcome
Anthropometric*		
Measure and plot on appropriate growth chart (for infants born prematurely, use corrected age until 2 years old): • Height (length) for age • Weight for age • Weight for height (length) or BMI • Head circumference (<3 years) Determine height-age (length)† , weight-age‡. Determine "ideal" weight§. Obtain and plot all previous anthropometric data that are available. Compare current data to previous measurements. Calculate incremental weight gain, linear growth and head circumference growth since last measurements obtained and compare to reference data for age.[8]	If poor weight gain, evaluate need to: • increase energy and protein intake • alter oral/enteral intake to decrease stool or ostomy output. If weight gain is excessive, evaluate need to decrease energy intake (this is most often an issue with patients on parenteral nutrition). If poor linear or head growth with normal weight gains, refer to physician for medical evaluation of poor growth.	Weight for height (length) is between 10th and 75th percentiles. Child is gaining weight and growing at a normal rate for age (or corrected age).8
For children over 2 years of age measure: • Triceps skinfold • Mid upper arm circumference Calculate: • Arm muscle circumference • Arm fat area Compare to reference data for age.[9]	Use information from assessment of muscle and fat stores to help in assessment of nutritional status. Concerns: • low muscle and/or fat stores (often an indication of long term inadequate energy intake) • high fat stores with normal or low muscle stores (often an indication of overfeeding with TPN)	Muscle and fat stores within normal limits for age.
Biochemical See laboratory standards for normal values		
If receiving parenteral nutrition, see Chapter 11 and Appendix C. After parenteral nutrition is discontinued and enteral or oral feedings are sole source of nutrition, monitor serum levels every 1-3 months until levels are within normal limits. Once stable, measure every 6 to 12 months.	Dosages for specific vitamin and mineral supplements will vary depending on a child's age, size, and degree of deficiency; consult the child's physician and pharmacist for appropriate dosage of vitamin and/or mineral to treat deficiency.	Child's micronutrient needs are met and biochemical indicators are within normal limits.
Vitamin A	If deficiency, give additional vitamin A in water-soluble form.	Indicators of vitamin A status are within normal limits.
Vitamin E	If deficiency, give additional vitamin E in water-soluble form.	Indicators of vitamin E status are within normal limits.

continued...

Assessment (continued)	Intervention	Evaluation/Outcome
Vitamin D, calcium, phosphorus, alkaline phosphatase	If vitamin D deficiency, give high dose vitamin D. Make sure child is receiving at least DRI of calcium and phosphorus. Recheck serum vitamin D three months after high dose supplement is started. Once serum vitamin D is within normal limits (>30ng/mL); continue same high dose supplement level and continue to monitor serum 25(OH)D every 3 to 6 months.[10, 11, 13] If serum calcium or phosphorus are low, and vitamin D levels are normal, give supplements of calcium and/or phosphorus.	Indicators of vitamin D, calcium, phosphorus, and alkaline phosphatase are within normal limits.
Magnesium	If deficiency, give Magnesium Protein Complex	Indicators of magnesium are within normal limits.
Zinc	If deficiency, give zinc supplement	Indicators of zinc are within normal limits.
Monitor serum level of vitamin B12 every 3 to 12 months for the life of the patient (It can take years for vitamin B12 deficiency to develop.)	Once vitamin B12 levels are in the low-normal range, begin sublingual tablets, nasal spray, or intramuscular shots of vitamin B12 (cyanocobalamin). Monitor levels every 1 to 3 months. B12 deficiency may require treatment with intramuscular forms of B12, followed by long term supplementation using sublingual tablets or nasal spray to maintain normal levels within the body. Each patient will differ in their absorptive capacity and require individual treatment during close monitoring of serum levels every 3-12 months.	Indicators of vitamin B12 status are within normal limits.
Clinical		
Assess stool or ostomy output: • If stooling though anus, obtain information regarding number, size, and consistency of stools per day • If child has ileostomy or colostomy, obtain information regarding approximate volume of output each day	If stool output is high, consider: • need for medical evaluation • need for intravenous fluid (patients with SBS are at high risk for malabsorption and dehydration) Evaluate need to alter feeding: • may need to hold feeding advancement or reduce feeds slightly to previously tolerated volume • may need to consider change in feeding schedule (bolus versus continuous) • many need to consider a change in formula type If stool output is high, child may have bacterial overgrowth and need antibiotics. If severe and weight loss present or feeding not being tolerated, increased TPN may be necessary until bacterial overgrowth resolved. Child may have a viral gastroenteritis; therefore, needs careful management of fluid status until gastroenteritis resolves.	Stool or ostomy output is less than 40—50 mL/kg/day.2

Dietary		
Obtain diet history or 3 to 7 day food intake record, and analyze for energy and protein. If on parenteral nutrition, calculate energy and protein in parenteral nutrition solution, check contents of vitamin and mineral additives. (See Chapter 11 and Appendix C) Compare intake to DRI for age and to growth and weight gain. Consider: • energy and protein needs of infants and children with short bowel syndrome who are eating or receiving tube feedings are often greater than the DRI • infants and children on TPN will require less total energy than those fed enterally/orally because of losses in stool due to malabsorption and decreased needs for diet-induced thermogenesis. It essential to avoid over feeding of the child on TPN, as over feeding is associated with excessive fat deposition, and TPN-induced liver failure.10 • parenteral protein needs will be the same as for enteral/oral feeding.	Provide diet education on avoidance of fruit and added sugars to minimize stool losses. Adjust TPN based on the child's tolerated oral/enteral intake to meet recommendations for nutrient intake. Due to high energy needs of the orally feeding child because of malabsorption, offer education on frequent meals/snacks. Snacks may need to look more like meals. Also provide food suggestions on high energy, nutrient dense, and low added sugar foods.	Energy and protein intake is adequate to promote growth.

* For reference data and guidelines for taking accurate measurements, see Chapter 2.
† Height-age is the age at which the child's current height (or length) would be at the 50th percentile on the growth chart.
‡ Weight-age is the age at which the child's current weight would be at the 50th percentile.
§ Ideal weight is the weight that would place the child at the 50th percentile weight for height (or length).

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Chapter 19

NUTRITION INTERVENTIONS FOR CHILDREN WITH METABOLIC DISORDERS

Cristine Trahms, MS, RD, CD, FADA Updated by Beth Ogata, MS, RD, CSP, CD

Most inherited metabolic disorders are associated with severe illness that often appears soon after birth. Neurologic impairment and intellectual disability may occur. However, effective newborn screening programs and advanced diagnostic techniques and treatment modalities have greatly improved the outcome for many of these infants (1,2).

Advances in newborn screening technology offer the potential for earlier diagnosis, prevention of neurologic crisis, and improved intellectual and physical outcomes.
When tandem mass spectrometry techniques are used in newborn screening laboratories, infants with a broader range of metabolic disorders can be identified, and identification can be earlier than ever before.

The goal of treatment for inborn errors of metabolism is to strive for correction of the biochemical abnormality. The approach to treatment for each disorder depends on the enzyme(s) affected and the metabolic consequences of that effect (2,3).
Without effective nutrition therapy many children with metabolic disorders would not survive infancy or would have severe cognitive and physical problems. Outcome of treatment for metabolic disorders is variable and depends on early diagnosis and intensive intervention.

For children with metabolic disorders, appropriate growth reflects the achievement of metabolic balance. In addition to a limited energy intake, inadequate weight gain may reflect a chronic elevation in ammonia levels or chronic acidosis. If growth
and development are to proceed normally, energy and all required nutrients must be provided in adequate amounts. At the same time, controlling the biochemical abnormality necessitates the restriction of nutrients specific to the disorder to the requirement level (3,4).

Most children with metabolic disorders require the restriction of one or more
nutrients or dietary components. These restrictions are specific to each disorder

and include, for example, the restriction of specific amino acids or total protein, fatty acids, simple sugars, or total carbohydrate. In general, the strategies for treatment focus on reducing the negative impact of the affected enzyme and normalizing biochemical status. These goals can be achieved by using one or more dietary modification or intervention strategies, depending on the disorder:

- reduce the substrate
- provide the product(s)
- supplement co-factors
- enhance elimination of excess nitrogen

The protein and amino acid restrictions require the critical assessment of protein and energy intakes; particular attention must be paid to the protein-energy ratio of these prescribed diets. See Table 19-1 for nutritional restrictions and modifications for selected metabolic disorders.

The nutrient needs of each individual must be carefully considered and the dietary prescription based on the individual genetic and biochemical requirements for nutrients. If the specific nutrient needs of an individual are ignored or
misunderstood, intellectual disability, metabolic crisis, growth failure, neurologic crisis, organ damage, or death may occur.

For many metabolic disorders, especially those involving amino acid metabolism, use of a specialized semi-synthetic formula (medical food) is needed to correct the metabolic imbalances caused by the disorder and meet the nutrient requirements for growth, maintenance, and activity. The formulas are generally supplemented with small amounts of high biological value (HBV) protein to supply the restricted amino acid(s) to the requirement level. These formulas can provide 75-80% of the total protein intake for the individual. Nitrogen-free foods are often needed to provide an appropriate energy intake, such as low protein pasta, bread, and other baked goods (3,4).

Maintaining metabolic balance requires frequent and intensive monitoring of biochemical parameters specific to the disorder and those indicative of normal nutrition status. The goal is to achieve biochemical levels at or near the normal range. Laboratory parameters that are frequently monitored include:

- plasma amino acids
- hematological status
- protein status
- electrolytes
- blood lipid level
- ammonia

Table 19-2 describes general biochemical monitoring guidelines for selected disorders.

Other considerations in management of metabolic disorders include monitoring (3-9):

- Hydration status: Dehydration in children with metabolic disorders often causes severe metabolic imbalance. Fluid intake and requirements must be carefully monitored. Constipation is also of medical significance.
- Illness: The "usual childhood illnesses" often cause the child with a metabolic disorder to lose metabolic balance and become seriously ill. Frequently, children require hospitalization and the administration of intravenous fluids to prevent metabolic "crisis." During infection or illness that results in catabolism, protein- containing formula is often refused. Continued administration of some form of energy and fluids assists in rehabilitation.
- Feeding: Some children who have neurological difficulties develop oral-motor problems that interfere with the provision of adequate nourishment. A hyperactive gag reflex is a frequent problem. Some providers use nasogastric or gastrostomy tubes as a feeding adjunct to prevent metabolic crisis.

The crucial role of nutrition support cannot be disputed in the treatment of these disorders. Effective treatment requires the expertise of a team, generally comprised of a geneticist, registered dietitian nutritionist (RDN), genetic counselor, psychologist, and neurologist. This team of experts is familiar with the nuances of current treatment for metabolic disorders and will incorporate new treatment innovations as they are deemed appropriate. However, the complex nutritional and medical management of these children cannot occur without the follow-up and support of the community teams. Communication between the team at the tertiary center, the community teams, and the family is crucial.

Table 19-1: Some Metabolic Disorders Amenable to Nutritional Therapy*

Disorder	Enzyme: missing or inactive	Biochemical features	Nutritional treatment	Adjunct treatment
Amino Acid Disorders				
Phenylketonuria	Phenylalanine hydroxylase	↑↑ blood phe ↑ phenylketones in urine Progressive, severe ID, which can be prevented by early treatment	Food: low protein Specialized formula: without phenylalanine, supplement tyrosine	sapropterin dihydrochloride (Kuvan®) pegvaliase-pqpz (Palynqiz™)
Tyrosinemia type 1	Fumaryl-acetoacetate hydrolase	Vomiting; acidosis; diarrhea; FTT; hepatomegaly; rickets ↑ blood/urine tyrosine, methionine; ↑ urine parahydroxy derivatives of tyrosine Liver cancer	Food: low protein Specialized formula: without tyrosine, phenylalanine, and methionine	nitisinone (Orfadin® and Nityr™)
Homocystinuria	Cystathionine synthase or similar	Detached retinas; thromboembolic and cardiac disease; mild to moderate ID; bone abnormalities; fair hair, skin; ↑ methionine, homocysteine	Food: low protein Specialized formula: without methionine, supplement L-cystine	Betaine (Cystadane®), folate, vitamin B12, Some respond to vitamin B6 supplements
Urea Cycle Disorders				
Ornithine transcarbamylase deficiency	Ornithine transcarbamylase	Vomiting; seizures; sometimes coma → death. Survivors usually have ID, ↑ plasma ammonia, glutamine	Food: low protein Formula: without non-essential amino acids	L-carnitine, phenylbutyrate,‡ L-citrulline, L-arginine Hemodialysis or peritoneal dialysis during acute episodes
Citrullinemia	Argininosuccinate synthetase	↑ plasma citrulline, ammonia, alanine Neonatal: vomiting; seizures; coma → death *Infantile:* vomiting; seizures; progressive developmental delay	Food: low protein Formula: without non-essential amino acids	L-carnitine, phenylbutyrate,‡ L-arginine

Carbamoyl phosphate synthetase 1 deficiency	Carbamoyl phosphate synthetase	Vomiting; seizures; sometimes coma → death Survivors usually have ID, ↑ plasma ammonia, glutamine	Food: low protein Formula: without non-essential amino acids	L-carnitine, phenylbutyrate,‡ L-citrulline, L-arginine Hemodialysis or peritoneal dialysis during acute episodes
Argininosuccinic aciduria	Argininosuccinate lyase	↑ plasma argininosuccinc acid, citrulline, ammonia Neonatal: hypotonia; seizures Subacute: vomiting; FTT, progressive developmental delay	Food: low protein Specialized formula: lower protein without non-essential amino acids	L-carnitine, phenylbutyrate,‡ L-arginine
Arginase deficiency	Arginase	Periodic vomiting; seizures; coma Progressive spastic diplegia, developmental delay ↑ arginine, ammonia with protein intake	Food: low protein Specialized formula: lower protein without non-essential amino acids	L-carnitine, phenylbutyrate ‡
Organic Acid Disorders				
Methylmalonic aciduria	Methylmalonyl-CoA mutase, or similar	Metabolic acidosis; vomiting; seizures; coma; chronic kidney disease; pancreatitis ↑ organic acid, ammonia levels	Food: low protein Specialized formula: lower protein without isoleucine, methionine, threonine, valine	L-carnitine, vitamin B12 IV fluids, bicarbonate during acute episodes
Propionic aciduria	Propionyl-CoA carboxylase, or similar	Poor feeding; vomiting; hypotonia; metabolic acidosis; seizures; coma; heart abnormalities ↑ ammonia, propionic acid; ↑ methylcitric acid in urine	Food: low protein Specialized formula: lower protein without isoleucine, methionine, threonine, valine	L-carnitine, biotin IV fluids, bicarbonate during acute episodes
Isovaleric acidemia	Isovaleryl-CoA dehydrogenase	Poor feeding; lethargy; seizures; metabolic acidosis; ↑ ammonia; isovaleric acid	Food: low protein Specialized formula: without leucine	L-carnitine, L-glycine

continued...

Disorder (continued)	Enzyme: missing or inactive	Biochemical features	Nutritional treatment	Adjunct treatment
Ketone utilization disorder	2-methylacetoacetyl-CoA-thiolase, or similar	Vomiting; dehydration; metabolic ketoacidosis	Food: low protein Specialized formula: without isoleucine Avoid fasting, emphasize high complex carbohydrates	L-carnitine, bicitra
Biotinidase deficiency	Biotinidase, or similar	In infancy, seizures, hypotonia, rash, stridor apnea; in older children, also alopecia, ataxia, developmental delay, hearing loss		Supplemental oral biotin
Maple syrup urine disease	Branched chain ketoacid dehydrogenase complex	Seizures; acidosis ↑↑↑Plasma leucine, isoleucine, valine	Food: low protein Specialized formula: without leucine, isoleucine, valine	L-carnitine; some individuals respond to thiamin
Fatty Acid Oxidation Disorders				
Medium chain acyl-CoA dehydrogenase (MCAD) deficiency	Medium-chain acyl-CoA dehydrogenase	Vomiting, lethargy, hypoglycemia	Avoid fasting	?L-carnitine
Long chain acyl-CoA dehydrogenase (LCAD) deficiency	Long-chain acyl-CoA dehydrogenase	Vomiting, lethargy, hypoglycemia	Low fat, low long chain fatty acids, avoid fasting	MCT oil, ?L-carnitine
Long chain 3-hydroxy- acyl-CoA dehydrogenase (LCHAD) deficiency	Long-chain 3-hydroxy- acyl-CoA dehydrogenase	Vomiting, lethargy, hypoglycemia	Low fat, low long-chain fatty acids; avoid fasting	MCT oil, ?L-carnitine
Short chain acyl-CoA dehydrogenase (SCAD) deficiency	Short-chain acyl-CoA dehydrogenase	Vomiting, lethargy, hypoglycemia	Low fat, low short chain fatty acids, avoid fasting	MCT oil, ?L-carnitine
Very long chain acyl-CoA dehydrogenase (VLCAD) deficiency	Very-long-chain acyl-CoA dehydrogenase	Vomiting, lethargy, hypoglycemia	Low fat, low very long chain fatty acids, avoid fasting	MCT oil, ?L-carnitine

Carbohydrate Disorders				
Glycogen storage diseases (Type Ia)	Glucose-6-phosphatase	Profound hypoglycemia; hepatomegaly	Avoid fasting; high complex carbohydrates; low fructose, sucrose	Raw cornstarch Glycosade® (modified cornstarch) Supplemental iron
Hereditary fructose intolerance	Fructose-1-phosphate aldolase	Vomiting; hepatomegaly; hypoglycemia, FTT, renal tubular defects after fructose introduction ↑ blood/urine fructose after fructose feeding	No sucrose, fructose	
Galactosemia	Galactose-1-phosphate uridyl transferase	Vomiting; hepatomegaly; FTT; cataracts; ID; often, early sepsis ↑ urine/blood galactose	Eliminate lactose, low galactose, use soy protein isolate formula	

* Table adapted from reference 4

‡ Sodium phenylbutyrate and phenylacetate are chemicals administered to enhance waste ammonia excretion; other compounds producing the same effect are also used (e.g., Buphenyl®, Ravicti®).

Table 19-2: Biochemical Parameters to Monitor in Children with Metabolic Disorders*

Disorder	Parameter	Frequency
All disorders	Hematocrit, hemoglobin, ferritin	Twice per year, depending on age and health status
	Prealbumin	Twice per year, depending on age and health status
	Length or height, weight, weight-for-length or BMI, head circumference	At each clinic visit
	Intake of medical food and foods as contributors of critical nutrients	Monthly, at each clinic visit
	Protein, energy, fat, nutrients critical to specific metabolic disorder	Monthly, at each clinic visit
Phenylketonuria (PKU)	Plasma phenylalanine, tyrosine	Monthly, if child is well, more frequently if levels or intakes are unstable
Tyrosinemia	Plasma tyrosine, phenylalanine, methionine; nitisinone	Monthly, if child is well, more frequently if ill
Maple syrup urine disease (MSUD)	Plasma leucine, isoleucine, valine, alloisoleucine	Monthly, if child is well, more frequently if ill
Urea Cycle Disorders, e.g., Ornithine transcarbamylase deficiency (OTC), Carbamoyl phosphate synthetase deficiency (CPS), Argininosuccinic aciduria (ASA)	Plasma ammonia, electrolytes, plasma carnitine, plasma amino acids	At each clinic visit, more frequently if ill or illness is suspected
Organic acidemias, e.g., Methylmalonic aciduria, Propionic aciduria, Isovaleric aciduria	Urine organic acids, electrolytes, plasma carnitine, plasma amino acids	At each clinic visit, more frequently if illness is suspected
Ketone utilization disorder	Urine organic acids, plasma carnitine, electrolytes, serum ketones	If illness is suspected
Galactosemia	Galactose-1-phosphate	At each clinic visit

* Table adapted from reference 4. These are provided for general reference. See management guidelines for specific disorders and tailor monitoring to individual considering intake, growth, metabolic control, clinical symptoms, etc.
Abbreviations: ID – intellectual disability; FTT – failure to thrive/growth restriction

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Resource

National Coordinating Center for the Regional Genetics Networks (NCC). NCC Resource Repository. Online: https:/nccrcg.org/resources. Accessed January 20, 2020. This is a library of resources, including links to ACT Sheets and Algorithms (diagnostic tools) and a nationwide directory of clinical genetics services.

Chapter 20

KETOGENIC DIET FOR SEIZURE DISORDERS

Renee Williams, MEd, RD, CD and Elaine Cumbie, MA, RD, CDE, CD
Updated by Sarah Sullivan, MS, RDN, CD

The Ketogenic Diet (KD) is an established, effective nonpharmacologic treatment for many types of epilepsy. It is a high fat, adequate protein, and low carbohydrate diet. Ketosis occurs when the body's carbohydrate intake is limited and fat from the body or diet becomes the primary energy source for the body. The exact mechanism of the diet's anticonvulsant and antiepileptic effects is not known. However, it has been proposed that changes in cellular metabolism resulting in increased metabolic enzymes or altered brain pH decreases hyperexcitability, and hence a less epileptiform state. Another speculation is that the antiepileptic effect is exerted via neuroprotection. This may involve protection from free oxygen radicals or prevention of apoptosis. (1,2,3,4,5).

The beginning of the specific use of the KD dates to 1921 (5,6). The Mayo Clinic's 1921 article suggested that a high fat diet, adequate in protein and low in carbohydrate could mimic the effects of starvation, thus producing seizure control. The diet fell out of favor in 1938 when phenytoin (Dilantin) was discovered leading to the era of medication treatment for epilepsy (5). Revival of the KD began in the early 1990's with a Hollywood producer and writer, Jim Abrahams and his son Charlie. Charlie's seizures were refractory to medications and other treatments. Abrahams read about the KD, and took his son to Johns Hopkins Hospital to start the diet. Charlie's seizures stopped completely soon after starting the diet. Abrahams created the Charlie Foundation, published a book about the KD, created a Dateline TV program about the KD in 1994, and a made-for-television movie called, "First Do No Harm".

There are four KD approaches being practiced around the country (6). They are

1. Long chain triglyceride diet (classic or traditional KD diet)

2. Medium chain triglyceride (MCT) diet

3. Modified Atkins Diet (MAD)

4. Low Glycemic Index Diet

This chapter will focus on the traditional KD. The traditional KD is based on an exact ratio of fat to combined protein and carbohydrate. For example, a 4:1 ratio would have 4 grams of fat to every 1 gram of protein and carbohydrate combined. (1,4,7,8) At the 4:1 ratio, fat contributes 90% of calories. Protein and calorie requirements are established based on current DRI with the goal of meeting needs for growth. High fat foods are the foundation of meal plans, including foods such as heavy whipping cream, butter, oil, and mayonnaise. Fluid restriction does not play a role in seizure control, and is no longer recommended (8,9). Vitamins and minerals as well as fiber are inadequate in the KD, and must be supplemented (6,8).

The diet can be implemented on an outpatient or inpatient basis (6). The Johns Hopkins inpatient protocol for initiating and maintaining the KD has been gradually modified at Johns Hopkins and other centers, and is continually evolving. Some centers observe a fasting protocol of approximately 24 hours. Others observe an overnight fast with no food consumption after midnight (3). Extended fasting is not required to achieve ketosis and can cause more side effects. Two approaches are observed with respect to starting the diet. The first approach starts at a lower ratio with full calories, such as a 1:1 ratio. The ratio is to be increased based on patients' tolerance. The second approach is to provide the diet at a set ratio, such as a 3:1 or 4:1 ratio, but starting with 1/3rd calories, increasing daily until full calories are tolerated (10).

The outpatient approach does not involve fasting. The diet can be started at a lower ratio such as a 1:1 ratio then progressed in 3-5 day increments to a higher ratio. Hypoglycemia, acidosis, nausea, vomiting and lethargy can be minimized or avoided by implementing the diet according to this protocol.

The KD is particularly effective in controlling absence, atonic, myoclonic seizures and infantile spasms. However, it may be tried as a therapy with any child who has refractory seizures (11), especially after two anticonvulsants are used unsuccessfully. The diet can treat epilepsy effectively in infants through adulthood. Previously, it was thought that children younger than one year of age have more difficulty maintaining ketosis and experience a higher incidence of hypoglycemia. Historically, the diet has been felt to be most effective in children ages 2 to 5 years of age. Children and young adults alike have had success in controlling their seizures with the KD (9). The diet will control seizures in approximately one-third of children who have been unable to control them with medications. Of the remaining number of children, one-half will have some degree of improvement in their seizures and/or anticonvulsant medications reduced (2,4,8,11,12,14). Because of issues of non- compliance, older children may have more difficulty maintaining adequate ketosis and diet control. The motivation to control their seizures can be enough to keep compliance adequate (11,14).

The KD is best initiated under the supervision of an experienced KD team. A team is best defined as a physician or epileptologist, nurse, registered dietitian nutritionist (RDN) and social worker who all have experience with the KD (8,11,15). A pharmacist can also be a valuable part of the

KD team. The KD is not an exact science, and since all children are different, an individualized approach based on current best practices is advised. Much is learned through experience, and by adjusting the diet as needed.

RDN's are charged with the job of translating the science of the diet into a palatable form. The advent of the Ketocalculator has decreased the time involved in calculating menus. Increased formula options such as Ketocal (Nutricia North America) and Ketovie (Ajinomoto Cambrooke), and specialty low carbohydrate foods have expanded options for the diet.

The availability of the team, especially the RDN during the diet initiation and the first three months of the diet are crucial to the success of the diet. It is difficult for families to absorb all of the information during the hospital admission, and they need continuing guidance from the KD team in order to address their concerns.

The remainder of this chapter presents guidelines for nutrition assessment, intervention, and evaluation/outcome for monitoring a child on the ketogenic diet.

Table 20-1: Ketogenic Diet for Seizure Disorders

Assessment	Intervention	Evaluation/Outcome
Anthropometric*		
Measure and plot on appropriate growth chart: • Height or length for age • Weight for age • Weight for height (or length) or BMI • Head circumference (under 2 years) Identify ideal body weight (IBW).† Compare all current measurements to reference data for age and to previous measurements.	Adjust recommendations for energy intake, based on growth, activity level. Goal is to allow for child to continue to grow according to current weight/height channels • To ensure accurate calories, obtain three-day food record. Evaluate average daily calorie intake. • Caloric adjustments are made slowly, and not combined with a ratio adjustment.	Child's growth continues appropriately. Weight maintenance for the first full year on the diet (maintenance defined as staying within weight channel) Weight loss can result in hyperketosis and vomiting. Consistent energy intake is important. Weight gain can result in a loss of ketosis, resulting in loss of seizure control.
Biochemical		
MD to order the following serum lab tests (pre-diet and monthly for the first months). MD will determine schedule depending on stability of the child.		
Total cholesterol Triglycerides	• Modification of fat sources may be needed if increased levels do not decline or stabilize, shifting from saturated fat to more polyunsaturated fat • Adding omega fatty acids (fish oil) is an effective therapy to decrease serum triglycerides. • Increase soluble fiber • If above interventions unsuccessful, consider reducing ratio	Normal fasting triglyceride and cholesterol levels
Free carnitine Esterified carnitine Most children become deficient at some point on the diet, especially at the higher ratios. Deficiency can decrease effectiveness of the diet to control seizures. Prolonged use of anticonvulsants such as valproic acid can deplete carnitine.	Begin supplementation with L-carnitine if needed.[4]	Normal free carnitine levels

Electrolytes	Monitor for severe dehydration. Recommend using table salt to taste when cooking. Consider use of potassium chloride (salt substitute) to maintain normal serum levels if needed. Bicarbonate of soda (baking soda) may be used to normalize low serum carbon dioxide. Low serum carbon dioxide can contribute to vomiting, especially during the initiation phase.	Normal electrolyte levels
Creatinine BUN Magnesium Phosphorus Calcium Protein Vitamins E, D Vitamins A /Retinol binding protein	Minor abnormalities may not need to be corrected. Consult with ketogenic diet team. • Maintain normal vitamin and mineral intake by using appropriate supplemental vitamin with iron • Antiepileptic medications (AED's) interact with vitamin D; generally any child on an AED needs a vitamin D supplement. Vitamin D low in almost all cases at start of diet • Meet DRIs for all vitamins and minerals, particularly Calcium and B vitamins as the diet is low in these.	Serum creatinine, BUN, magnesium, phosphorus, calcium, protein levels, Vitamins A, E, D and retinol binding protein within normal limits.
Albumin, Prealbumin	If albumin is low, increase protein intake.	Albumin levels within normal limits.
AED (antiepileptic medication) levels	Some anticonvulsants can have an increase in medication level when a state of ketosis is present.	• MD to determine if decrease in anticonvulsants is indicated
Aspartate aminotransferase (AST) Alanine aminotransferase (ALT)	Fat absorption/metabolism may be decreased in liver disease - KD can alter liver function due to high load of fat • High AST and ALT may indicate the need to decrease ratio	Normal AST and ALT levels
Complete blood count	Close physician supervision is necessary to evaluate. • Provide iron supplement if anemic. • Elevated hemoglobin and hematocrit may indicate fluid deficit; obtain fluid intake history, determine possible causes	• Maintain normal hematocrit and hemoglobin. • Maintain adequate hydration, based on fluid needs per weight
Urine ketone levels • Checked at same time twice daily, useful at the beginning of the diet. • After initial start, only a good indicator of dehydration or hyperketosis in the context of decreased oral intake of fluids or food.	Ideally, urine ketones should be checked every morning and afternoon. If ketones are checked just once per day, then afternoon or evening is necessary. Caregivers should log ketone levels several times a week along with seizure activity to help evaluate the success of the diet.	• AM Ketones: consistent level, individualized for each child • PM Ketones: consistent level, individualized for each child • Discontinue once child has stabilized on the diet

continued...

Assessment (continued)	Intervention	Evaluation/Outcome
Betahydroxybutyric Acid Large ketone body found in serum, considered the most reliable method for determining level of ketosis		Consistent range depending on child's response to the KD.
Selenium	If deficient, start with 40 mcg Selenium, then recheck serum values Increase dose as needed until serum levels are normal.	Maintain selenium within normal serum limits.
Zinc	If deficient, supplement with 15 mg zinc, then recheck serum values. Increase dose if needed until serum levels are normal.	Maintain zinc within normal serum limits.
Clinical: Monitor side effects of diet		
Acute symptoms: Lethargy	May be seen within the first couple of weeks of diet initiation.	Transient lethargy is normal. Normal activity returns.
Acidosis	An expected side effect In the presence of low carbon dioxide, consider starting sodium bicarbonate or baking soda	pH within normal range
Nausea/vomiting	May be a sign of hyperketosis. Give 15-30mL juice if symptoms of high ketones above usual range, including nausea and vomiting. Encourage patient to take all of daily fluid allowance. Dehydration can occur quickly with moderate to severe vomiting. If no improvement within 24 hours or vomiting is severe, contact MD.	Ketones within normal limits based on child's normal values. Nausea and vomiting are diminished.
Hypoglycemia	Trend of lower blood sugars on the diet is common, but s ymptomatic hypoglycemia can occur when initiating the diet or during illness. Symptoms include: • Pallor and fatigue • Nausea • Excessive drowsiness • Diaphoresis • Confusion Symptomatic hypoglycemia should be treated.11 Give 15-30 mL of juice.	Blood glucose levels remain within acceptable limits without symptoms of hypoglycemia.

Chronic Effects		
Hyperlipidemia	No long-term cardiovascular side effects are known. If triglyceride levels are constantly rising and do not subside, the risk of complications versus the benefit of the diet must be considered. Phenobarbitol can increase serum triglycerides.	Serum triglyceride and cholesterol in acceptable range. See biochemical section
Vitamin or mineral deficiency	Provide carbohydrate-free multivitamin/ mineral and calcium supplement if formula does not meet DRIs. The diet is inadequate for most vitamins and minerals. Anticonvulsant medication-nutrient interactions are common. See Chapter 5 for specific assessment and intervention guidelines.	Ensure adequate amounts of vitamins and minerals.
Growth	Some slowing in linear growth may occur on the diet.	Growth should be plotted at each follow-up visit and provide adequate protein.
Constipation • Determine pre-diet bowel pattern • Intervene if child is constipated before start of diet	Due to the lack of fiber in the diet, many children require intervention. Miralax, Dulcolax, Colace, glycerine suppository, fiber and milk of magnesia may be used. Use of lower carbohydrate fruits and vegetables (10% fruits and group A vegetables)‡ should be encouraged in order to maximize the serving size of fiber-containing foods.	Bowel movements should be achieved at least every 1-2 days.
Kidney stones Increased risk if family history or use of AED carbonic anhydrase inhibitor (CA-I)	Evaluation by a renal specialist is preferred. Continuation of the diet may be possible with increase in fluid intake. Consider potassium citrate prophylatically. Calcium intake greater than the DRI is discouraged.[2,4,8]	
Noncompliance	This is the most common problem. It is more prevalent in older children and poorly organized families. Decreased betahydroxybutyrate and increased seizures are typical. Diet calculations should be rechecked for miscalculation or excessive energy, including addition of protein or carbohydrate which results in ratio change. Possible errors in food preparation should be discussed. Not giving all of the fat in meal plan can also cause decreased ketosis. Caregivers must be encouraged to be "sleuths" in looking for possible mistakes or extra carbohydrate in the diet (medications, toothpaste, and "sugar-free" beverages).	Strict adherence to the diet for at least 3 months for adequate evaluation of diet success on seizure control. Continued strict adherence to the diet is necessary to maintain seizure control.
Dietary		
Obtain diet history and/or 3 day food record, including all food preferences.	Review intake.	Intake is appropriate, with consideration of nutrients discussed below.

continued...

Assessment (continued)	Intervention	Evaluation/Outcome
Review all medications and nutritional supplements currently used. Assess amount of carbohydrate provided by supplements and medications. Ketocalculator contains the carbohydrate content of many common supplements and medications.	Medications and nutritional supplements should be converted to lowest carbohydrate-containing form. In general, no chew tabs or liquids should be used and tablets are preferred lowest-carbohydrate form[11].	Review medication list at each clinic visit. Make sure nothing has been used or added that is not on the approved list. Breakthrough seizures can occur.[16]
Assess energy needs	Daily energy goal—using IBW, a starting point of 75-100% DRI for age is appropriate unless current energy intake is significantly greater or less than the DRI[4,11]. Adjust recommendations for energy intake, based on rate of weight gain. Excessive weight gain can negate the effects of the diet. Slow weight gain may be appropriate if ketosis is achieved.	Slow weight gain, maintain BMI percentile, stable betahydroxybutyrate (no extreme fluctuations up or down.)
Assess protein needs	Daily protein goal—protein should be kept at DRI per kilogram IBW if possible, with a minimum 1.0 g/kg for children less than 7 years of age and a minimum 0.8 g/kg for children 7 years and older. Meeting adequate protein levels and desired ratio can be difficult in children with low energy needs[4,11].	Normal protein status is maintained.
Assess fluid needs	Daily fluid goal per Holliday-Segar equation. Adjust as needed per labs, risk of kidney stones, or hydration status. Educate caregivers on signs and symptoms of dehydration[4,11].	Maintain adequate hydration status. High BUN or hematocrit may indicate mild dehydration.
Develop diet prescription and meal plans	Establish meal schedule—3 meals/day is standard, but smaller, more frequent meals may be needed. If tube-fed, determine tube-feeding regimen. Both bolus and continuous regimens can be achieved[11].	Maintain steady ketosis by eating evenly spaced meals and consistent ratio at each meal.

Develop diet prescription and meal plans 1. Caloric needs based on three-day diet history and growth history 2. Dietary preferences based on preference survey	Establish ratio (defined as grams of fat to grams of protein and carbohydrate combined). Consult with KD Epileptologist to determine starting ratio, and best plan for KD start. This will be dependent on age of child, and overall caloric need. Some programs start with a 24-hour fast or longer, some only fast after midnight the day before the diet start. Initiation options: 1. Consistent ratio, increase calories over three days, 1/3, 2/3, then full calories. 2. Progressive ratio ramp-up, 1:1, 2:1, 3:1 for example. 3. Outpatient diet initiation program - start with the lowest ratio, then ramp up the ratio every 3-7 days until the desired ratio is achieved. Lower ratios may be necessary for children with very low energy needs in order to maintain adequate protein in the diet.	Ketogenic diet ratio sufficient to maintain consistently high ketosis, and optimum seizure control. Betahydroxybutyrate within consistent range.
	Recommend using the Charlie Foundation Keto Diet calculator. Computer programs give more accurate information and are faster. Families can be trained to calculate meal plans when appropriate.	Families should be given at least 10-15 meal plans to initiate diet and taught how to calculate meals. Caloric needs are met, providing weight maintenance or slight gain to maintain BMI percentile.

continued...

Assessment (continued)	Intervention	Evaluation/Outcome
Determine education needs	Ketogenic Diet education—caregivers come with different levels of understanding and preparation regarding the diet. Education program should include: • History of the diet • Theoretical basis • Expectations of the diet • Possible complications • Expectations of the program • Resources needed • Ketone testing • How to handle illness • Importance of compliance • Monitoring requirements • Nutritional guidelines—energy, protein, carbohydrate, fat, fluids, vitamins, and minerals • Meal planning—eating consistently, using a gram scale, careful measuring, recipe suggestions, and meal preparation Give caregivers as much written information as possible in an organized format for home reference. Provide adequate education, likely in outpatient setting.	Caregivers plan, prepare, and measure meals with 100% accuracy. Ketosis is stable as demonstrated by stable betahydroxybutyrate. Weight gain consistent with defined parameters.
Assess appropriate time and regimen for weaning the diet	Recommended minimum of 3 months on the diet, and often discontinued after 2 years. However, wean should be individualized based on the child's needs as determined by family and medical team, including the KD Epileptologist. Diet is to be weaned slowly, typically over 2-3 months by gradual reduction in ratio. If seizures increase, the diet can be reversed. Not all children have total seizure control, or become medication-free.	Monitor seizure control during and after weaning from the diet.

* For reference data and guidelines for taking accurate measurements, see Chapter 2.
† Ideal weight is the weight that would place the child at the 50th percentile for weight for height (or length).
‡ See Freeman JM. The Epilepsy Diet Treatment: An Introduction to the Ketogenic Diet, 2nd Edition, p.39 for more information.

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Additional Resources

Charlie Foundation

Charlie Foundation for Ketogenic Therapies
515 Ocean Ave., #602N
Santa Monica, CA 90402
www.charliefoundation.org
Ketogenic Diet Calculator: http://www.ketodietcalculator.com

The Charlie Foundation is an education resource for the ketogenic diet. The Foundation was established by Jim and Nancy Abrahams, whose son Charlie had become seizure free on the ketogenic diet.

Epilepsy Foundation of America

3540 Crain Highway
Suite 675
Bowie, MD 20716
https://www.epilepsy.com

This is a national organization that provides information about epilepsy to professionals and persons with epilepsy. They provide community services, public education, federal and local advocacy, seizure first aid training, and research funding into new treatments and therapies.

Nutricia North America

https://nutricia-na.com
http://www.myketocal.com

This is the website sponsored by Nutricia North America and offers resources for both patients, families, and healthcare professionals. Parents can find resources at https://www.myketocal.com) and professionals can find them at Nutricia Learning Center. Resources include information about the diet, formula, recipes and more.

Books

The Ketogenic Diet: A Treatment for Children and Others with Epilepsy

Freeman, J. et al.
Demos Medical Publishing, 2006. 386 Park Ave. South, Suite 210 New York, NY 10076 800.532.8663

This is a good basic book for professionals and families alike who are interested in learning more about the diet.. To read an excerpt from the book, go to the Amazon.com website, under department, select books, and type in Ketogenic Diet.

Keto Kid: Helping Your Child Succeed on the Ketogenic Diet

Snyder, D, Freeman, J. et al. Demos Medical Publishing, 2006. 386 Park Ave. South, Suite 210 New York, NY 10076 800.532.8663

This book was written by a family physician whose child is on the Ketogenic Diet. It provides recipes, as well as helpful hints for families to promote success while on the diet. To read an excerpt from the book, go to the Amazon.com website, under department, select books, and type in Ketogenic Diet.

Ketogenic Diet Therapies for Epilepsy and Other Conditions, Seventh edition

Kossoff, E. et al.
demosHEALTH, 2020
11 W. 42nd St.
New York, NY 10036

This book goes through a little bit of everything you need to know about ketogenic diets.

The Keto Cookbook: Innovative Delicious Meals for Staying on the Ketogenic Diet

Martenz, D, Cramp, L
demosHEALTH, 2011
11 Ws. 42nd St.
New York, NY 10036

This book is a collection of recipes written by a mom of a child on the diet, so all of these recipes are kid tested. There are some helpful cooking hints in here and cooking tools that would be helpful on a ketogenic diet. It also goes over some of the trickier things like batch cooking and emergencies.

Chapter 21

NUTRITION INTERVENTIONS FOR AUTISM SPECTRUM DISORDER

Betty Lucas, MPH, RD, CD
Updated 2022 by Sharon Feucht, MA, RDN, CD

Autism spectrum disorder (ASD) is a complex neurodevelopmental condition involving persistent challenges with social communication/interactions and restricted, repetitive patterns of behavior. Diagnostic criteria, present before the age of 3 years, are described (1). While autism is considered a lifelong disorder, the degree of impairment in functioning because of these challenges varies among individuals with autism (2). According to the CDC's Autism and Developmental Disabilities Monitoring Network's surveillance program, the prevalence of ASD was about 1 in 54 children in 2016, which is 2.7 times higher than the prevalence reported in 2000 (3). It occurs more in males than females, and approximately 33 percent of those diagnosed with ASD have intellectual disability.

Children with ASD[1] reflect a wide range of behaviors and severity. Interventions are individualized and commonly include speech and behavior therapies to improve social interactions, behavior, and communication. Children diagnosed with ASD may have other identified neurodevelopmental disabilities (i.e., fragile X syndrome, Down syndrome, Attention Deficit/Hyperactivity Disorder), learning disabilities, anxiety disorders, or speech and language disorders (4). In addition, other medical conditions can impact the health and/or behavior of those with ASD and their families. These conditions include seizures, sleep disorders, gastrointestinal (GI) disorders, feeding disorders, and obesity.

Although the specific etiology of ASD is not known, causes include genetics and environmental factors. At this time, families with one child diagnosed with ASD are at increased risk of having other children with ASD. Other factors increasing the risk of ASD include age of parents, extreme prematurity, low birth weight, and multiple gestation pregnancies. Certain compounds in the environment, such as organophosphates and nitrogen dioxide, may increase risk at a population level (4). Future research will yield more information on the causes of ASD.

ASD symptoms can be identified by primary care providers through prescribed developmental screening and/or with a specific screening tool for ASD completed at 18 and 24 months of age

[1] While person-first language for autism (e.g., child with autism) is often used in health care and research settings, many autistic individuals prefer the use of identity-first language (e.g., autistic child), and there is no universal consensus within the community. When working with individual patients and families, defer to their preference on the language used to describe the relationship between an autism diagnosis and the individual.

(4, 5). While the goal is to identify children with ASD early in life so that intervention services can begin, some children may not be identified until older. Resources are also available for parents/caregivers to review if they have concerns about their child's development (6). If screening is positive or shows a child is at risk, a comprehensive diagnostic evaluation is indicated, along with referral to services such as early intervention or special education (1).

ASD is diagnosed by behavioral features in two domains: social communication/interactions and restricted/repetitive patterns of behaviors. Behaviors include lack of communication and interaction with others, inflexibility of behavior, difficulties tolerating changes in routine, and sensory hypersensitivity. These behavioral features can impact all aspects of a child's life, including nutrition and feeding. Table 21-1 provides examples of behaviors that may interfere with nutrition and feeding.

Table 21-1 Examples of Behaviors of Children with Autism Spectrum Disorder that Interfere with Nutrition and Feeding

- Difficulty in communicating hunger cues leading to random eating schedules or grazing
- Difficulty in transitions to meals/snacks or changes in mealtime routines which may lead to tantrums/food refusal
- Increased sensory sensitivity with restricted intake due to smell, taste, or texture of foods. May reject foods from one or more food groups
- Rigidity in accepting foods based on color, shape, or appearance
- Rigidity in eating locations, tableware, and food placement on plates
- May be easily distracted at meals/snacks
- Decreased acceptance of foods over time
- Refusal of "new" foods

During a diagnostic evaluation for ASD, questions about nutrition and feeding should be reviewed. In addition, routine medical visits should screen for feeding issues in children diagnosed with ASD (7). Feeding difficulties and/or impaired nutrient intake in individual children with ASD can vary from mild to severe. When feeding/nutrition concerns are noted, referral to an individual health care professional and/or an interdisciplinary feeding team, including a Registered Dietitian Nutritionist (RDN), should be made (7).

Nutrition and Feeding

Breast/chestfeeding is recommended for all infants. A 58% decrease in the risk of ASD was seen in children if they were ever breast/chestfed (8). For those children exclusively breast/chestfed, a 76% decrease in risk for ASD was noted (8). More studies are needed to understand this breast/chestfeeding connection and the relationship to ASD diagnosis.

Little research is available to evaluate the introduction of complementary foods to infants later diagnosed with ASD. Early referrals for feeding evaluations can support families who have concerns about how or what their infant is eating. Following the evaluation, if issues are identified, intervention can start in a timelier manner and be positive for both the family and the child.

As children move beyond infancy the appearance of feeding concerns and challenges with food acceptance may appear in those diagnosed with ASD. Parents report "picky" eating in their child with ASD; this behavior is more persistent than the transient picky eating typically seen in young children. The term "selective eating" is often used and has been defined as measurable aspects of food refusal, limited food repertoire, and high frequency, single food intake (9).

Children with ASD are reported to have a five-fold increase in feeding difficulties compared to typically developing children (7). Up to 70% of children with ASD may experience atypical eating behaviors compared to children with other disorders (10). In addition to selective eating behaviors, atypical feeding behavior can include hypersensitivity to food textures, pocketing food, and/or pica (10). The severity of feeding and nutrition concerns can vary greatly in children diagnosed with ASD. A small study reported that children assessed at age 6 and again at age 13 did decrease overall food refusal; however, teens did not increase the number of different foods eaten (9). Many support interventions for feeding difficulties early in life for those with ASD.

Children may narrow their food acceptances, and some with ASD may accept fewer than 10-15 items, reject foods from one or more food groups, only accept certain brands or methods of preparation, refuse new foods, and may reject a meal or snack if foods and/or presentation vary from their preferred pattern. Negative behaviors, including tantrums, may develop around mealtimes. Eventually, parents may respond by giving their child foods they know he/she will eat. This leads to different and sometimes separate meals for the child, and if behavior is an issue, less participation in family mealtime. This can also lead to inadequate or excess nutrient or energy intake.

Children with ASD were reported to have significantly lower intakes of protein, calcium, phosphorus, selenium, vitamin D, thiamine, riboflavin, and vitamin B12 but higher intakes of polyunsaturated fatty acids (PUFA) and vitamin E when compared to typically developing peers (11). When the amounts the children with ASD consumed were compared to the appropriate Dietary Reference Intake recommendations, the risk of low intake appeared only in calcium and vitamin D and showed adequate intakes of the other mentioned nutrients (11).

A subset of children with ASD may have severe food selectivity. Parents have reported omission of vegetables and fruits from their children's diets. In addition, these children were at risk for inadequate intakes of five or more nutrients including vitamin D, fiber, vitamin E, and calcium. These children were not at risk for compromised growth or obesity (12).

Some children with ASD become so selective in their eating that a diagnosis is made of avoidant/restrictive food intake disorder (ARFID). ARFID may be diagnosed when the child consistently does not meet nutritional or energy needs, leading to significant weight loss/failure to grow, significant nutrient deficiency (e.g., vitamins A, C, D, B12, and thiamine), and/or dependence on oral supplements or tube feedings (13).

Feeding Behaviors

The persistent selective eating and related behaviors that limit the food intake of a child with ASD can be very challenging to all who work with the child. In children with ASD, correlates of feeding difficulties have been reviewed. An individual child's sensory processing profile, challenging mealtime behaviors including rigidity, and the presence of constipation can contribute to increased feeding challenges (14). As noted earlier, if parents report concerns with nutrition and feeding for their child with ASD, a referral to the RDN and/or a team of other health care professionals (e.g., medical providers, feeding therapists, behavior therapists, the RDN, and others) is appropriate.

Interdisciplinary team evaluations address the broad areas listed below. With a team evaluation, the RDN role may be to focus more closely on nutrition, meal patterns, and growth, but also cover other aspects of assessment to compare findings with team members (see Chapter 1 for specific components of nutrition assessment) (15).

- Physiological: e.g., sensory processing, ability to recognize hunger and satiety, sleep
- Medical: e.g., GI issues (see below), food allergies, oral-motor abilities, nutrient deficiencies, medications
- Psychological-Behavioral: e.g., family food interactions, repetitive/restricted interests, fear/anxiety, previous interventions leading to negative interaction with foods
- Oral Motor: e.g., swallowing dysfunction, oral-motor and feeding delays, limited experiences

Some children with ASD eat a variety of foods and do not exhibit food selectivity. Families may need guidance from the RDN about meal/snack schedules or specific foods to incorporate into the child's diet based on the assessment. Additional therapies may be needed, but overall, the child eats a variety of foods, is growing appropriately, and exhibits typical food selectivity.

Other children with ASD may be diagnosed with moderate food selectivity. Research has been reported about parent education programs. These programs are guided by an interdisciplinary team, including the RDN. They feature education for families over a period of 10-11 weeks and include behavior modification techniques, nutrition planning, meal structure, and introduction/incorporation of new foods (16, 17).

For children with severe feeding concerns and/or ARFID, referral to an interdisciplinary team providing direct intervention may be required. An intensive, multidisciplinary feeding intervention can increase dietary variety and nutrient intake, as well as improve mealtime behaviors (18).

Interdisciplinary feeding teams, including the RDN and a behavior therapist, can be difficult to locate in a community, and services may be limited to children with the most restricted and nutrient-limited diets. The RDN may need to reach out to the other providers for an individual

child to collaborate with nutrition and feeding interventions if a designated "team" is not available. With the goal of diagnosing ASD earlier in young children, it is hoped that referrals for selective eating can be made early to avoid development of more serious feeding and nutrition concerns.

Feeding and nutrition Interventions can be incorporated into a child's educational goals, such as an Individualized Family Service Plan (IFSP) or Individualized Education Program (IEP). This requires coordination between the school, family, and the RDN and/or the feeding team. Figure 21-1 at the end of the chapter is a handout for families that describes eating behaviors in children with ASD and offers guidance for caregivers.

Gastrointestinal Issues/Microbiota and ASD

Gastrointestinal (GI) issues, such as constipation, diarrhea, and gastroesophageal reflux disease, are reported more often in children diagnosed with ASD (19). In young children ages 2-5 years, those diagnosed with ASD were two times more likely to have parent reported gastrointestinal symptoms than children with other developmental delays and three times more likely than children in the general population. Those with ASD may be unable to communicate about or share where pain/discomfort is located due to social communication and/or sensory processing issues. Problems may be indicated through increasing negative behaviors, sleep issues, anxiety, and/or aggression (20). If a family reports recent changes in behavior, sleep, etc. or decreases in food intake, exploration of these concerns should occur and/or referral for a GI evaluation may be needed (21). Screening tools using parent reports for commonly identified GI issues in children have been researched (22).

A possible gut-brain connection in those diagnosed with ASD has been widely suggested with differences in gut bacteria noted between those with and without ASD. The role of the gut microbiota and whether alterations affect GI status and/or behavior are not well understood in those with ASD. It is unclear if these differences are a result of the diagnosis, a reflection of dietary intake, both, or due to other reasons. To date, research on the use of commercial sources of probiotics, prebiotics, a combination of both, or fecal transplants is limited and inconclusive (23). The use of these products is not supported at this time as a therapeutic intervention in ASD. Families may need counseling if products are used to understand their expectations of the intervention. Offering a variety of food sources of both probiotics and prebiotics is appropriate for all individuals. Acceptance of a variety of these foods may be difficult for those with ASD and increased food selectivity.

Other Nutrition Related Concerns in Children with ASD

Growth

Many children diagnosed with ASD have growth parameters within normal limits. However, individual children can be at risk for under- or overnutrition (24). Youth with ASD are at an increased risk of obesity compared to their typically developing peers (25). Individuals with ASD

may experience barriers to engaging in active leisure or organized sports, have repetitive eating patterns including energy-dense foods, and are more likely to be prescribed medications, such as atypical neuroleptics (or antipsychotic medications) and anticonvulsants, that often contribute to excessive weight gain. Increased screen time and sleep disorders may further predispose them to obesity. Primary care providers should monitor a child's age-specific BMI percentile in the context of health supervision care and address modifiable risk factors through anticipatory guidance and/or referral to appropriate programs.

A multidisciplinary intervention for weight management has been reported for children 5-12 years of age (26). Two virtual nutrition education programs have been described for adolescents with ASD (27, 28). A review of weight management interventions for youth with ASD has been published (29).

Physical Activity

Children 4-7 years old with ASD have been reported to engage in the same levels of moderate to vigorous physical activity as typically developing children. (30). Decreased physical activity has been associated with increased age in children and adolescents diagnosed with ASD (31). Adolescents with ASD were less likely to meet guidelines for physical activity of one hour per day and exceeded screen time recommendations of greater than two hours per day (32). Children 6-11 years old with ASD were less likely to meet both physical activity and screen time recommendations. In addition, they were less likely to meet guidelines for daily sleep duration (32). Review of a child's physical activity during a nutrition assessment can guide intervention suggestions. Opportunities for activity can be identified for families, education settings, and community groups.

Pica

Some children and youth with ASD put nonfood items in their mouths beyond early childhood (4). This behavior may result of sensory differences or a behavior that persists from early childhood. If a feeding and/or nutrition assessment indicates pica, follow-up is needed to ensure items are not harmful and that there is not a medical reason for this behavior. The child's iron and lead status should be evaluated. Interventions can identify more appropriate chewing substitutes for a child needing oral input for soothing. Behavioral interventions and/or removal of items from the child's environment may be required along with supervision of the child's surroundings.

Dental Health

Children with ASD have been reported to have a higher risk of dental caries, which may be related to the types of foods consumed, grazing throughout the day, and/or food pouching in the mouth due to swallowing issues. In addition, increases in dental injuries due to pica, tongue thrusting, bruxism, and/or self-injurious behaviors have been reported. There is an increased prevalence of gingivitis among children with ASD, which may be related to irregular brushing or inability to brush effectively. Sensory issues can impact the effectiveness of daily dental care (33).

As with other children, anticipatory guidance should include attention to dental hygiene and fluoride use, if appropriate, from a young age. The RDN can explore the child's meal/snack patterns and types of items eaten to evaluate if the child is at increased risk for dental caries. Referral to a dental home is recommended for all children based on risk assessment, as early as 6 months of age, 6 months after the first tooth erupts, and no later than 12 months of age (34). The American Academy of Pediatric Dentistry provides information for parents related to dental health and their children and youth with special health care needs (35).

Complementary and/or Alternative Medicine (CAM) Interventions

Families with a child diagnosed with ASD report using a variety of CAM interventions, including special diets and supplements. The plethora of information available to families is often presented with sophisticated marketing, testimonials, and claims which may prompt many parents to adopt dietary changes and supplementation regimens for their children (36). Analysis of research about the most common alternative nutrition interventions has, in general, not supported the use of supplements and/or diets as a recommended therapy to relieve GI symptoms and/or improve behavioral symptoms for those with ASD. Many studies have been small in size, limited in duration, and often do not reflect a rigorous research design (37). Nutrition assessments should include a review of supplements, special diets, and any other complementary and alternative products used. A complete nutrition assessment may indicate the need for specific special diets and/or supplementation that can benefit an individual child.

Understanding the nature and scope of alternative therapies is important in providing the best quality, family-centered care. RDNs and other health professionals must be able to evaluate information about alternative nutrition therapies for ASD and use evidence-based practice. Families who are searching for effective treatments for their children are often unable to discriminate between anecdotal reports and research-based scientific evidence. For example, caregivers do not always realize that components of multivitamin mixtures can also be found in other supplements their child is taking. Families should be counseled on the tolerable upper limits (ULs) now set for most vitamins and minerals, and should be encouraged not to exceed those levels.

Open dialogue between the RDN and the family is critical, as is an understanding of the family's desire for an effective treatment. RDNs working with families using alternative nutrition therapies should:

- Verify all CAM use
- Review reasons for use of CAM based on child's unique feeding and nutrient information
- Share evidenced-based sources for information related to CAM
- Use evidenced-based information to educate caregivers about diet modifications, special diets, or elimination of food groups

See Table 21-2 below for commentary related to current research for the most common nutrition-related CAM (36, 37, 38, 39). Research is ongoing related to specific diet and nutrient interventions. Frequent review of evidenced-based research is suggested for the RDN working with families and their children diagnosed with ASD.

Table 21-2: State of the Evidence for Common Nutrition-Related CAM Therapies (36, 37, 38, 39)

Special Diets	
Gluten-free/casein-free diet (GFCF)	A subset of children with ASD may benefit. Attention should be paid to calcium and vitamin D if casein is eliminated and B vitamins if gluten is eliminated. Fiber sources may be low. It is difficult to delineate whether gluten or casein is affecting an individual child if both are eliminated. Long term treatment may be required to see a response. Adherence may be difficult for children with feeding issues. Recommend that families work with the RDN and other members of the health care team to monitor growth and nutrition status.
Specific carbohydrate diet (SCD)	Few studies and no symptom improvement noted; no randomized controlled trials (RCTs). Very restrictive diet; adherence may be difficult for children with feedings issues. If recommended, the family should work with the RDN and other members of the health care team to monitor growth and nutrition status.
Ketogenic diet	Some animal studies and very limited human studies may indicate behavior improvement. Very rigorous diet that may be unpalatable and cause nutrient deficiencies if not accepted by the child. Must be done under supervision of an interdisciplinary health care team, including the RDN.
Supplements	
Fatty acid supplements	Mixed and inconclusive results evaluating supplement sources of omega-3 fatty acids. Foods rich in beneficial fatty acids can be encouraged in the child's diet.
Vitamin A	Correction of vitamin A deficiencies did not affect ASD symptoms. Excessive use can be harmful.
Vitamin C	Recent studies in children have suggested a correlation between ASD and scurvy. Correction of dietary deficiency did not improve ASD symptoms.
Vitamin B_{12}	Few studies; early positive results; effects are inconclusive; additional research is needed.
B6 and magnesium	Historically, mixed results; more recently, a few reviews and meta analyses have reported no effect or insufficient evidence supporting an effect on ASD symptoms.
Vitamin Supplements advertised for ASD, i.e., Super-Nu-Thera®	No data supporting therapy; possible harm from excess intake if provided in amounts suggested.
Probiotics	See Chapter 6 and 7.
Other supplements, including digestive enzymes, levocarnitine, etc.	Limited research, and variable or no improvement in behaviors.

The remainder of this section presents guidelines for nutrition assessment, intervention, and evaluation/outcome for children with ASD.

Assessment	Intervention	Evaluation/Outcome
Anthropometric - See Chapter 2		
Measure and plot on appropriate growth chart: • Length/height for age • Weight for age • Weight for length or BMI • Head circumference (<2 years) Compare all current measurements to reference data and to previous measurements. Collect growth history from medical records and caregivers.	If underweight or overweight, work with family to modify child's energy intake by adjusting the type of foods offered, energy-density of foods, and frequency of eating, as well as adapting the feeding environment and addressing behavior problems as needed (see Dietary below).	Growth and weight-for-length or BMI are appropriate.
Clinical/Medical		
• Biochemical data, including screening for lead and iron exposure if pica behavior is present • Developmental history • History of GI disease or infections • Constipation, diarrhea, reflux • Medications used • Oral health care, i.e. toothbrushing, dental care	Address any dietary interventions for constipation or diarrhea (see Chapters 6 and 7). Determine any medication-nutrient interactions, and adjust any nutrient supplements and/or food choices, consulting with primary care provider as needed (see Chapter 5). Provide education re: food and snacks that are less cariogenic; encourage regular dental care and exams.	No symptoms of GI disorders. Any medication-nutrient interactions are compensated by food or supplements. Child receives daily oral care and routine dental exams.
• Determine usual activity level • Assess any constraints on physical activity due to safety or other issues • Assess amount of screen time (mobile devices, computer, TV, video games, etc.)	• Support daily, safe physical activity, e.g., preschool children should be active throughout the day. For children 6-17 years of age, recommend 60 minutes of mostly aerobic activity per day; encourage muscle strengthening and weight-bearing activities three days per week as part of the recommended 60 minutes daily. Sports appropriate to age, development, and need for supervision. See Chapter 3. • Special Olympics program for children >8 years of age • Reduce screen time if greater than 2 hours per day or as appropriate for child's overall program.	Caregivers, educators, and others involved in child's daily care report appropriate and safe physical activity.

continued...

Assessment (continued)	Intervention	Evaluation/Outcome
Dietary - See Chapter 1		
Assess: • Typical food intake, timing of snacks and meals, and eating environment • Methods of preparation and preferences, e.g., brands, presentation, color, shape, temperature • Early feeding development history • Feeding Concerns – this information may be obtained from assessment by another qualified health care professional if part of a team • Food aversions, refusals • Use of food as a reward • Feeding behaviors -- current and past • Previous dietary interventions • Use of special diets, i.e., GFCF, SCD, and nutrients at risk, i.e., calcium, Vit D • Vitamin and/or mineral supplements, herbals, etc. • Other complementary and alternative medicine (CAM), i.e., enzymes, probiotics Obtain a three- to seven-day food record. Determine usual energy and nutrient intake	• If child is overweight or underweight, adjust energy intake by altering portion sizes, increasing or decreasing snacks, and changing beverage volume and/or energy-density. Plan should accommodate child's food limitations and preferences as much as possible. See Chapter 13. • For micronutrient inadequacies, collaborate with family to find alternative food sources that might be acceptable, e.g.., suggest other calcium-fortified food and beverage sources for those with dairy allergy. • Provide vitamin-mineral supplements if needed; work with family regarding acceptable form (chewable, liquid, crushed), color, taste, etc. Treat supplements as "medicine" or use a behavioral approach to increase compliance. • Provide family education and counseling regarding ULs for supplements, evidence-based effectiveness, and any dangers of toxicity or negative side effects. • Discourage use of food as a reward in all environments; work with family and professionals to identify other reinforcers. • Support family meals and regularly-scheduled meals and snacks.	Subsequent re-evaluation of food records indicate appropriate energy and nutrient intake from a variety of foods. Caregivers report dietary practices consistent with nutrition care plan.
Family/Social		
Determine: • Family's concerns about child's nutrition and feeding, and current goals for change • Child's behaviors that limit or alter food intake • Previous attempts to improve diet and intake, and their outcomes • Use of CAM therapies and family's expectations	Nutrition counseling for caregivers, educators, and others to set small goals with the expectation that progress will be slow (6): • Food exposures should be step-wise, i.e., child tolerates, smells or touches food, but does not eat initially. See Figure 21-1. • Refer for feeding therapy by OT, SLP, and/or behavior specialist at early intervention, school, or other program (with consultation from RDN) Consider group feeding therapy for preschool children.	Child makes slow, but consistent progress with food acceptance and improvement in feeding behaviors. Periodic re-assessment of family's concerns and goals.

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Resources

Autism Speaks

https://www.autismspeaks.org/

Autism Speaks is committed to providing the autism community with resources to meet each person and family's unique needs throughout every stage of life. There are resources and commentaries at this site on a wide variety of topics related to ASD. In the information section there is a link to Health and Wellness topics related to ASD including nutrition, feeding, fitness, sleep, constipation and medications.

Specific nutrition and feeding guides and toolkits are available at:

https://www.autismspeaks.org/nutrition

Autism Society of America

http://www.autism-society.org/

Information about ASD is available along with direction to state and/or community affiliates. This group works on public policy and offers resources on living with ASD for families/children and adults. There are state and/or community affiliates of the Autism Society of America. In Washington the state affiliate and community groups can be reached at:

http://autismsocietyofwa.org/v2/

Centers for Disease Control and Prevention
https://www.cdc.gov/ncbddd/autism/index.html
CDC provides a variety of information related to ASD.

Washington State Department of Health (DOH)https://www.doh.wa.gov/youandyourfamily/illnessanddisease/autism/statewideresources
WA DOH provides state focused information and resources at this website.

Nutrition Focus Newsletter –
https://nutritionnetworkwa.org/resources/nutrition-focus/
This newsletter features nutrition issues of children and youth with special health care needs and their families. The newsletter is designed for use by health care providers and others who serve these children and youth in a community or ambulatory setting. Two recent issues discussed ASD:

- Novak P and Perez J. Nutrition Assessment and Treatment of Autism Spectrum Disorders. *Nutrition Focus Newsletter*. University of Washington. 2017;32(4):1-14.
- Novak P and Perez J. Nutrition and Autism Spectrum Disorders – Part 2; Treating Obesity in ASD. *Nutrition Focus Newsletter*. University of Washington. 2017;32(5):1-10.

Maternal and Child Health Digital Library
https://www.mchlibrary.org/professionals/autism.php

National Institutes of Health
Office of Dietary Supplements
http://ods.od.nih.gov/

National Institutes of Health
National Center for Complimentary and Alternative Medicine
https://www.nccih.nih.gov/health/autism

National Institutes of Mental Health
Autism Spectrum Disorders
https://www.nimh.nih.gov/health/topics/autism-spectrum-disorders-asd

Medline Plus: Complementary and Alternative Therapies
http://www.nlm.nih.gov/medlineplus/complementaryandalternativetherapies.html

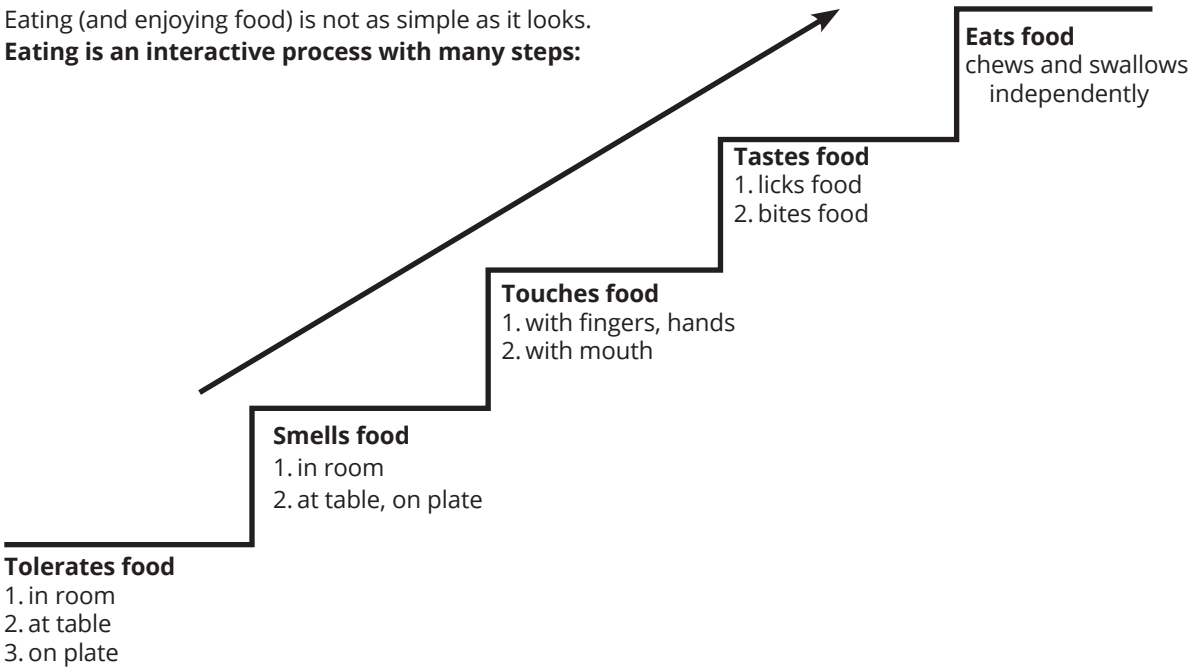
Fig. 21-1 AUTISM, NUTRITION, AND PICKY EATING…

- Some common behaviors in children with autism can cause problems around food and eating. (Your child may or may not have these behaviors.)

BEHAVIOR	HOW IT MIGHT RELATE TO CHILDREN'S EATING
Hypersensitivity to texture, smell, taste	Refuses to eat foods with specific textures, smells, tastes
Need for routine	Refuses to eat food that looks "different"… or is in a new bowl … or is at a different place at the table
Overstimulated or overwhelmed by the environment	Distracted from meal and does not eat

- Typically developing children may need 10 or more exposures to a new food before they will eat it.

…children with autism who are especially sensitive to tastes, textures, smells, or new things may need even more exposures!



Adapted from "Steps to Eating" by Kay Toomey, Ph.D., Denver CO

- ➢ It is natural for children to refuse to eat some foods at some times. This is one way to show independence and to make decisions.
- ➢ Picky eating can be a nutrition risk when:
- One or more food groups is excluded from a child's food pattern
- "Not enough" food leads to weight loss or lack of weight gain
- Fights over eating (or not eating) put a strain on parent-child relationships

WHAT MAY WORK **..but not in the long run**	WHY THIS IS NOT IDEAL
"Bribing" your child to eat a food. *("If you eat your vegetables, you can have some candy.")*	Teaches your child that he/she shouldn't like vegetables as much as candy, because vegetables are less desirable
Forcing your child to *"take a bite"* or *"have at least one taste."*	Creates negative associations with food and eating. Takes control away from your child.

Suggestions to help your child enjoy new foods

Avoid overwhelming your child with too many changes:

1. Keep mealtimes constant. Use the same plates and utensils. Eat at the same place and at the same time.
2. Offer small servings of a few (2-3) foods at one time...avoid offering TOO MUCH FOOD and TOO MANY CHOICES.
3. Offer new foods along with foods your child already likes to eat.

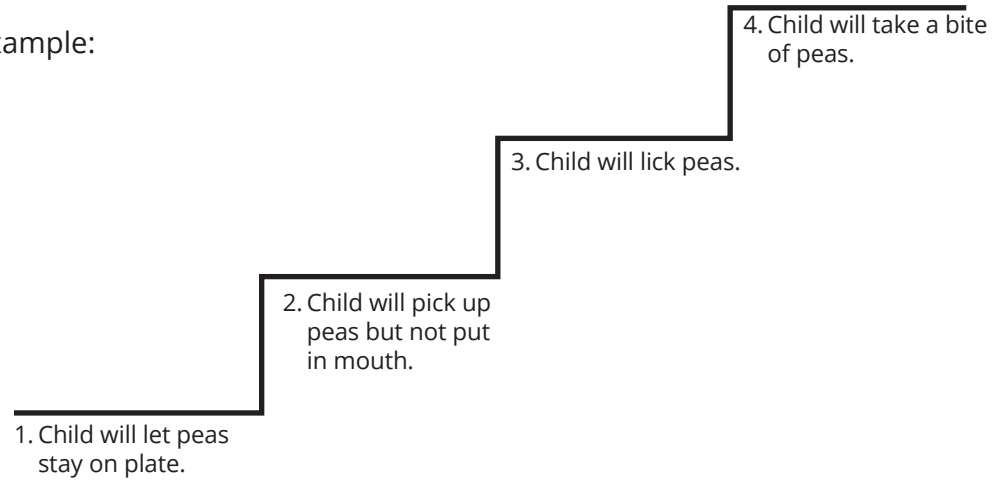
Introduce foods in forms that are similar to foods your child already eats, and make changes gradually.

For example, if your child eats crackers, but not sandwiches:

- First, offer sandwiches made with crackers
- Then, offer sandwiches made on toast
- Finally, offer sandwiches made with bread

Expect slow changes. Follow your child through the steps of the process... set realistic goals.

For example:



Ask your child's teacher for help.

1. Pick one "goal food" to offer at snack time and/or lunch. Offer this same food at home.
2. Use non-food reinforcers. Using food as a reinforcer teaches your child to value this food—and can teach your child not to value other foods.
3. Incorporate eating behaviors into your child's token reward system—a token for a "goal behavior," such as leaving peas on plate. Remember NOT to use food as the end reward

Developed by Beth Ogata, MS, RD and Betty Lucas, MPH, RD
Center on Human Development and Disability, University of Washington, Seattle, WA
September 1999

Appendix A

NUTRITION SCREENING

PEACH * Survey

Agency: _____ Date: _____

Child's Name: _____ Date of Birth: _____

Address: _____ Phone #: _____

_____

Please circle YES or NO for each question as it applies to your child.

Does your child have a health problem (do **not** include colds or flu)? If yes, what is it?	YES NO	1
Is your child: Small for age? ___ Too thin? ___ Too heavy? ___ (If you check any of the above, please circle YES)	YES NO	3
Does your child have feeding problems? If yes, what are they?	YES NO	3
Is your child's appetite a problem? If yes, describe:	YES NO	1
Is your child on a special diet? If yes, what type of diet?	YES NO	2
Does your child take medicine for a health problem (Do **not** include vitamins, iron, or fluoride)? Name of medicine(s):	YES NO	1
Does your child have food allergies? If yes, to what foods?	YES NO	1
Does your child use a feeding tube or other special feeding method? If yes, explain:	YES NO	4
Circle YES if your child does **not** eat any of these foods: Milk ___ Meats ___ Vegetables ___ Fruits ___ (Check all that apply)	YES NO	1
Circle YES if your child has problems with : Sucking ___ Swallowing ___ Chewing ___ Gagging ___ (Check all that apply)	YES NO	3
Circle YES if your child has problems with : Loose stools ___ Hard stools ___ Throwing Up ___ Spitting Up ___ (Check all that apply)	YES NO	3
Does your child eat clay, paint chips, dirt, or any other things that are not food? If yes, what?	YES NO	2
Does your child refuse to eat, throw food, or do other things that upset you at mealtime? If yes, explain:	YES NO	2
For infants **under 12 months who are bottle fed**: Does your child drink less than 3 (8-ounce) bottles of milk per day:	YES NO	1
For children **over 12 months**: (Check if applies and circle the YES) Is your child **not** using a cup? _____ Is your child **not** finger feeding? _____	YES NO	1
For children **over 18 months**: Does your child still take most liquids from a bottle?	YES NO	2
Circle YES if your child is **not** using a spoon?	YES NO	2

* Parent Eating and Nutrition Assessment for Children with Special Health Needs

Total = ____

The Parent Eating and Nutrition Assessment for Children with Special Health Needs (PEACH) survey.

Reprinted with permission from Campbell MK, Kelsey KS. The PEACH survey: a nutrition screening tool for use in early intervention programs. J Am Diet Assoc. 1994 Oct;94(10):1156-8. doi: 10.1016/0002-8223(94)91139-8. PMID: 7930323.

Appendix B

TECHNICAL ASPECTS OF ENTERAL FEEDING (TUBE FEEDING)

Annette Pederson, MS, RD, CD

Types of Enteral Feeding

The types of enteral feedings, or tube feedings, are named according to the feeding route used, the site where the feeding tube enters the body, and the point at which the formula is delivered: nasogastric, nasoduodenal, nasojejunal, gastrostomy, and jejunostomy. The decision as to which type of feeding to use is based on the expected duration of tube feeding as well as physiologic and patient-related factors. The types of tube feeding most commonly used are nasogastric and gastrostomy feedings.

Nasogastric Tube Feeding

The nasogastric (NG) tube is a soft tube that runs through the nose and into the stomach. Nasogastric feedings are typically used when tube feeding will be required for a short time, i.e., less than three months, although in some cases it can be used for several years. The major advantage of nasogastric, nasoduodenal, and nasojejunal feedings is that unlike gastrostomy or jejunostomy feeding, placement does not require surgery. Therefore, they can be started quickly and can be used either for short periods or intermittently with relatively low risk of complication. If the child is safe to feed orally, he can continue to practice feeding skills and improve oral intake.

Caregivers can learn how to place an NG tube at home and verify correct placement of the tube in the stomach by aspiration of stomach contents or listening for air bubbles in the stomach as recommended by their medical team. Nasal tubes need to be taped to the cheek, which can be irritating, and children may pull their tubes out. Taping nasal tubes is an art, and caregivers may need practice to find out what works best for them.

The disadvantages of NG feeding include nasal or esophageal irritation and discomfort (especially if used long-term), increased mucus secretion, and partial blockage of the nasal airways. Nasogastric feeding may contribute to recurrent otitis media and sinusitis. Two additional disadvantages are the possibility that the tube will perforate the esophagus or the stomach and the possibility that the tube will enter the trachea, delivering formula into the lungs. If formula enters the lungs, severe or fatal pneumonitis can result. Therefore, it is essential to confirm that the NG tube is in the stomach before feeding begins (1,2,3).

Gastrostomy Tube Feeding

A gastrostomy tube places food directly into the stomach. Gastrostomy feedings are preferred as they allow more flexibility with schedule and can mimic normal feeding schedules. These feeding tubes are well suited for long-term enteral feeding. Patient comfort with gastrostomy is an advantage over NG tubes. Gastrostomy tubes do not irritate nasal passages, the esophagus, or the trachea; cause facial skin irritation; or interfere with breathing. The mouth and throat are free for normal feeding if the child is safe to feed orally. There are low profile gastrostomy tubes that are easily hidden under a child's clothing, require less daily care, and interfere less with the child's movement. Gastrostomy tubes may be held in place with a balloon – a benefit where you can change the g-tube at home as the child grows or if the tube malfunctions. Non-balloon g-tubes cannot be replaced at home so the child will need to return to the hospital for replacement. A gastrostomy with a large-bore tube allows for a more viscous formula and thus, a lower risk of tube occlusion. The gastrostomy may be placed surgically. Another alternative is the percutaneous endoscopic gastrostomy (PEG), which may be done as an outpatient procedure.

G-tubes are sized using French sizes that indicate the diameter of the tube and the stoma length in cm, e.g., 18 FR 1.5 cm gtube. As the child grows, they may need to change g-tube sizes. Families should keep a spare feeding tube at home and have a plan to replace the tube if the tube becomes accidentally dislodged.

Disadvantages of gastrostomy feeding include the surgery or endoscopy required to place the tube, possible skin irritation or infection around the gastrostomy site, and a slight risk of intra-abdominal leakage resulting in peritonitis. A child with poor gastric emptying, severe reflux or vomiting, or at risk for aspiration may not be a good candidate for a gastric placed tube (1,2,3).

Jejunal Tube Feeding

Jejunal tubes can be placed surgically or via percutaneous endoscopy. Feeding directly into the jejunum (the middle section of the small intestine) is used for children who cannot use their upper gastrointestinal tract because of congenital anomalies, GI surgery, immature or inadequate gastric motility, severe gastric reflux, or a high risk of aspiration. The jejunal tube bypasses the stomach, decreasing the risk of gastric reflux and aspiration. There are Gastrostomy-Jejunal tubes (G-J) that are placed in the stomach with the feeding tube threaded into the intestine. The child is fed via the J-port, and the G-port is used for venting and medications. If safe to feed, the child can still eat by mouth.

However, even for children with gastric retention and a high risk of aspiration, there are disadvantages to jejunal feeding. Jejunal tubes passed from a gastrostomy to the jejunum and nasal-jejunal tubes are difficult to position and may dislodge or relocate; their position must be checked by x-ray. A jejunostomy reduces problems of tube position. Jejunal tube feedings usually require continuous drip feeding which results in limited patient mobility and may impair activities

of daily living. Finally, when compared to gastric feedings, they carry a greater risk of formula intolerance, which may lead to nausea, diarrhea, and cramps. Standard formulas may be given in the small intestine if tolerated, however, elemental or semi-elemental formulas may be required if the child demonstrates formula intolerance (2,4). Elemental formulas are more expensive (1,2).

Administration of Tube Feeding: Bolus and Continuous Drip

Tube feedings can be administered as bolus feedings, continuous drip feedings, or a combination of the two along with eating orally. The best is a combination of oral and tube feeding that fits into the child/family schedule. Many of the complications of tube feeding arise from improper administration of formula.

Bolus Feeding

Bolus feedings are defined amounts of formula or "meals" delivered four to eight times during the day. Typically, each feeding lasts about 15 to 30 minutes. The advantages of bolus feedings over continuous drip feedings are that bolus feedings are more similar to a normal feeding pattern, more convenient, and less expensive if a pump is not needed. Furthermore, bolus feedings allow freedom of movement, so the child is not tethered to a feeding bag. The parent/caregiver can hold and cuddle the child while feeds are given.

A disadvantage of bolus feedings is that they may be aspirated more easily than continuous drip feedings. For some children, bolus feedings may cause bloating, cramping, nausea, and diarrhea. It may not be practical to use bolus feedings with a child when the volume of formula a child needs is large or requires that the child be fed around the clock (1,3).

Continuous Drip Feeding

Continuous drip feedings are a specific amount of formula delivered during a specified time(s) during the day. Feeding around the clock is not recommended as this limits a child's mobility and may elevate insulin levels, contributing to hypoglycemia. It is common to use drip feedings for 8 to 10 hours overnight with children who cannot tolerate large volumes of formula. This also allows oral feeding to be used during the day. Either gravity drip or infusion pumps deliver continuous drip feeding. The infusion pump is a better method of delivery than gravity drip. The flow rate of gravity drip may be inconsistent and needs to be checked frequently. A child may start out with continuous drip feedings and as tolerance improves, graduate to bolus feedings or a combination of the two.

Continuous feeding may be better tolerated than bolus feeding by children who are sensitive to volume, are at high risk for aspiration, or have gastroesophageal reflux. Continuous feeding

can be administered at night, so it will not interfere with daytime activities. When feedings are delivered continuously, stool output is reduced, a consideration for the child with chronic diarrhea. Continuous infusions of elemental formula have been successful in managing infants with necrotizing enterocolitis, short bowel syndrome, intractable diarrhea, and Crohn's disease.

A disadvantage of continuous feeding is that the child is "tied" to the feeding equipment during the infusion, although feedings can be scheduled at night and during naptime. However, there are smaller and lighter pumps available to make life easier for the family. Continuous feeding is more expensive because of the cost of supplies, including the pump. Management of a pump and its maintenance may be difficult for some families. Finally, a child's medication needs to be considered, as continuous feeding may interfere with serum concentration of some medications (1,2,3).

Equipment for Tube Feedings

Feeding Tubes

When choosing a feeding tube, the following factors should be considered: the patient's age and size, the viscosity of the formula to be used, and the possible need for a pump.

Nasogastric tubes

For nasogastric feeding, the smallest bore tube in a soft material will minimize the child's discomfort. Large-bore tubes partially block the airways, may interfere with the function of the gastroesophageal sphincter, and may irritate the nose and throat.
Tubes size 8 French or smaller are usually used for children. The size refers to the outside diameter of the tube; one French unit equals 0.33 mm. Tubes this small cannot accommodate thick or viscous feedings, e.g., homemade blenderized formula or commercial formula containing fiber (1,2,5,6).

Most tubes are made of polyurethane or silicone, both of which remain soft and flexible over time. These tubes are usually weighted at the end for easier insertion. More flexible tubes are difficult to place without using a stylet. Non-weighted tubes may be displaced during gagging, vomiting, or coughing spells; however, they are used regularly and without difficulty for intermittent feedings in newborns.

Gastrostomy tubes

Skin-level gastrostomy feeding devices, such as the Bard® button gastrostomy or the MIC-KEY® button gastrostomy are available. They allow feeding tubes to be attached only when the child is being fed. These devices are easily hidden under a child's clothes without tubing that extends from the stomach. They may be placed surgically or endoscopically in the stomach

wall or after a gastrostomy tube has been placed and the stoma site is well established. Some gastrostomy tubes have extensions that "lock" into place, so the child is less likely to become disconnected while feeding, e.g., MIC-KEY®. Some parents prefer the flatter fitting tube such as the EndoVive® and Bard®. There are many more gastrostomy tubes available and parents can ask their physician or infusion company for more information. Families should check with their home supply company to determine how many gastrostomy tubes are provided every 6 months. If the child has the balloon gastrostomy tube, e.g., MIC-KEY® or EndoVive®, the parents need to be instructed on what to do if the tube comes out or how to replace it with a back up gastrostomy tube.

Pumps

A pump can be helpful in providing a consistent amount of formula to the child, which may improve tolerance. Pumps are designed to be accurate within 10% of the set rate, so determining the accuracy of what the patient is actually getting is important. With small infants, a syringe bolus pump will provide more accuracy with a small volume (7).

There are many different enteral infusion pumps, varying in complexity, flow rate, language, and cost. Pumps can be rented or purchased from a medical supply company. For long-term use, it may be less expensive to buy a pump. When a patient owns a pump, they will be responsible for its maintenance, while the medical supply company is responsible for repair and maintenance when renting a pump.

When deciding which type of pump to use, there are various considerations: availability, accuracy, cost, and ease of maintenance. A lightweight, battery operated enteral feeding pump, such as the Zevex Infinity®, will allow the child to attend school or go on outings with formula and equipment fitting neatly into a small backpack. If the child is fed at night only, an ambulatory pump may not be needed.

For some families, pumps do not fit into their life. Syringe bolus feeds require less equipment and instruction time, and alarms do not beep in the night.

Feeding Sets/Extensions

Many of the pumps require specific feeding sets, including a container for the formula and tubing to connect the formula container to the feeding tube. Reuse of feeding sets can help minimize the cost.

After use, the formula container, drip chamber, and tubing should be carefully cleaned. Rinse first with cold water, followed by hot soapy water and then rinse thoroughly to remove the formula residue which can cause bacterial contamination. Using a vinegar or bleach rinse before the final water rinse is helpful.

ENFit is the new standard for enteral tube feeding connectors. This replaces Legacy or pre-ENFit connectors, decreasing the risk of misconnections. Tubing misconnections can have severe consequences in hospital and home settings.

The family needs to know how many extensions, feeding bags, and other supplies their insurance company or home infusion company will provide. Usually, one feeding bag/day and one connecting tube per week is provided. Families may choose to save money and reuse feeding bags and tubing.

Additional Equipment

To give the child more mobility during continuous feeding, the feeding set can be hung on an IV pole and connected to a long length of tubing. Alternatives are to hang the feeding set on a hook above the bed or crib, or on the back of a wheel chair or bedpost. When traveling by car, the pump can be placed on the back seat with the feeding set hanging from the clothes hook. Miscellaneous supplies include syringes, gauze, barrier cream, and tape.

Nutritional Considerations

In order to determine energy and nutrient needs, nutritional status should be assessed before tube feeding is started. Table B-1 outlines the requirements of typical infants and children for water, energy, and protein; requirements are based on the Dietary Reference Intakes (DRIs). These requirements are useful in formulating tube feedings for children and youth with special health care needs, as long as conditions that may alter the child's nutrition needs are taken into account. For example, cardiopulmonary stress may increase energy needs, while decreasing tolerance to fluid volume; infection or the stress from surgery may increase both energy and protein needs; and certain medications may increase the requirement for specific vitamins or minerals. On the other hand, immobility tends to decrease energy needs, and if formula intake is limited calorically, there may be deficiencies in protein, vitamins, and minerals.

Table B-1: Water, Energy and Protein Requirements for Children (8)

	Water		Energy		Protein	
	cc/kg	cc/lb	kcal/kg	kcal/lb	gm/kg	gm/lb
STEP 1 For first 10 kg or 22 lbs, provide:	100	45	105	48	2.0	0.9
STEP 2 For second 10 kg or 22 lbs, provide:	50	24	50	23	0.9	0.4
STEP 3 For weight over 20 kg or 44 lbs, add to the amount above an additional:	20	10	20	9	0.3	0.2

Energy

The only way to accurately evaluate an individual child's energy needs is to regularly monitor weight gain, growth, and actual energy intake. If energy intake is inadequate, weight gain will be poor. If energy intake is excessive, weight gain will be higher than that desired for linear growth. Factors that may change energy needs include illness, increased seizures, surgery, increase in therapy, return to school, or changes in medication.

Children who have been chronically underweight while on oral feedings often gain excessive weight when tube feedings are initiated. For these children, two factors may come into play: oral-motor problems that interfere with adequate energy intake by means of oral feeding, and energy needs that are lower than expected. Cases such as these illustrate the necessity of routinely monitoring weight and energy intake in children who are tube fed, especially after the tube feeding is initiated.

Older children with delayed growth due to inadequate intake may have delayed puberty. With adequate energy provided by tube feeding along with increased body fat, they may begin to experience pubertal growth and body changes into their twenties. These changes need to be assessed when determining energy needs, so weight gain is appropriate.

Fluid and Electrolytes

Water must be provided in sufficient quantities to replace fluid losses and allow for normal metabolism. Fluid requirements depend on the following variables: urine output, sweating, vomiting, fever, stool pattern, environment, renal disease, cardiac anomalies, tracheostomies, and medications. Constant drooling also contributes to fluid losses. Water requirements can be estimated using Table B-1 as long as the above variables are considered. Indications that fluid intake is inadequate include constipation, decreased urine output, strong smelling or dark urine, crying without tears, dry lips and skin, sunken eyes, and weight loss. Symptoms of fluid overload

include rapid weight gain, puffy appearance, and rapid or uncomfortable breathing.

Patients who rely on tube feedings as their sole source of nutrients are at risk for electrolyte imbalances, which may result in serious medical complications, e.g., hyponatremia, hypernatremia, hypokalemia, hyperkalemia, dehydration, and cardiac arrhythmias. Sodium, potassium, and chloride status should be evaluated regularly (1,4). The DRIs/RDAs provide guidelines for a safe and adequate intake of electrolytes.

Vitamins and Minerals

To determine vitamin and mineral needs, the DRIs/RDAs for age can be used as a base, unless the child's growth is markedly delayed. For the child with growth delay, the DRIs/RDAs for height-age can be used. Children with inadequate energy intakes, decreased absorption, and increased energy needs should be evaluated for supplemental vitamins and minerals.

Vitamin and mineral requirements can be altered by medications (See Chapter 5). Other variables to consider are disease, previous medical and dietary history, and biochemical parameters.

Minerals that require special attention are calcium, phosphorus and iron; these are usually not adequate in commercial tube feeding formulas at the energy levels required by many children. Supplemental vitamins and minerals can be given with feedings in the form of a liquid multivitamin with iron or a crushed chewable multivitamin/mineral tablet appropriate for age. Children on long-term enteral support are at risk for trace mineral deficiencies. The risk of developing micronutrient deficiencies increases with frequent vomiting or gastrointestinal disturbances. Children on long-term tube feedings need to be evaluated for fluoride intake and may need to be supplemented. This will require a prescription from the child's physician or dentist.

Children with cystic fibrosis or anomalies of the distal ileum and ileocecal valve may fail to absorb fat-soluble vitamins or to reabsorb bile salts (see Chapters 15 and 17).

Formulas

A wide variety of commercial formulas are available for tube feeding. See Appendix F for information about the various commercial nutrition products and formulas. Formula selection should be individualized, based on nutrient and fluid requirements, gastrointestinal function, and reimbursement coverage.

Standard infant formulas (or specialized infant formulas, if needed) can be given via tube. Pediatric enteral formulas are designed specifically to meet the nutrient requirements of most children 1-10 years of age. These formulas are complete and balanced, i.e., about 1000-1300 ml will meet 100%

of the DRI/RDA for vitamins and minerals. These formulas are isotonic and easily tolerated by most children. There are disease-specific formulas that may or may not meet pediatric nutrition needs. An adult formula may be used for the older child, however the adult formula may not meet the child's vitamin and mineral needs. The protein and fiber content of adult formulas are higher than a child may require, so special attention is needed to make sure a child receives adequate fluid. One formula may not meet all the needs of a child; two or more formulas may be needed to individualize a specific nutrition plan. When selecting an appropriate formula, the factors to consider include the following:

- Age and medical condition
- Nutrient requirements and goals
- History of food intolerance or allergy
- Intestinal function
- Route of delivery
- Formula characteristics, e.g., osmolality, viscosity, nutrient content, convenience, and cost
- Availability of product

The osmolality of a formula has a direct influence on the gastrointestinal (GI) side effects that occur with enteral feeding. Osmolality refers to the concentration of osmotically active particles per kilogram solution of formula, expressed as mOsm/ kg. The osmolality of a formula is affected by the concentration of amino acids, carbohydrates and electrolytes. Formula with a higher osmolality than that of normal body fluids produces an osmotic effect in the stomach and small intestine; this hyperosmolality draws water into the GI tract to dilute the concentration of the formula. An influx of water into the GI tract may cause diarrhea, nausea, cramping, and distention. Isotonic formulas are designed to prevent these problems. The osmolality of full-strength isotonic formulas is similar to the osmolality of normal
body fluids, approximately 300 mOsm/kg water.

Sometimes parents feel that because enteral formulas are not solid food, they are not feeding their child enough. Parents need reassurance that you will help their child to not feel hungry and their nutrient needs can be met by formula alone.(2,6)

Complete or Standard Formulas

Complete or standard formulas are nutritionally complete and made of complex proteins, fats, carbohydrates, vitamins, and minerals. Complete formulas are designed for patients who have normal digestion but cannot consume adequate energy and nutrients orally. The advantages of complete formulas are that they have low osmolalities and are lactose-free, easy to use, and sterile. Some complete formulas have added fiber.

Elemental Formulas

Elemental formulas are “predigested” formulas made from amino acids or hydrolyzed protein, simple carbohydrates, and fat in the form of medium chain triglycerides and essential fatty acids. They contain all the essential vitamins and minerals. The major advantages of elemental formulas are that little or no digestion is required, stool volume is low, and the stimulation of bile and pancreatic secretions is minimal. Elemental formulas are hyperosmolar however and may cause cramping and osmotic diarrhea if infused too rapidly. They are more expensive than standard formulas and offer no advantage to a child whose gut is intact. Clinical indications for the use of elemental formulas include short bowel syndrome, malabsorption syndromes, delayed gastric emptying or gastroesophageal reflux, inflammatory bowel disease, gastrointestinal fistulas, cystic fibrosis, and nonspecific malabsorptive states.

Homemade Blended Formulas (9)

Families often ask about making their own blenderized feedings. This may be a less expensive alternative to formula. Many families want to nourish their child with the same foods the family is eating. Commercial formulas made from blenderized table foods (e.g., chicken, fruits, and vegetables) are available but may not provide complete, balanced nutrition for children; they will need to be evaluated by an RDN (see Appendix F). Families may want to use a combination of blenderized feedings for home and commercial formula for school, travel, etc.

Home blended formulas can be less expensive but more time intensive to prepare. Families need to be aware of their child’s dietary needs and restrictions. The RDN can help support the family by monitoring growth, hydration, and formula tolerance, and can analyze the formula to ensure optimal growth and nutrient intake. Parents should understand food safety to prevent contamination of the formula (10). When using a home blended formula, supplements may be necessary to meet nutrient requirements, along with additional water to meet fluid needs. A larger sized gtube >14 FR is helpful for the thicker formula.

A meal plan can be created for the child. USDA My Plate is a meal plan that mimics family meals, providing a variety of foods to meet the child’s needs. It is a free resource available online and via smartphone app. A meal plan can also be made using the exchange method to meet specific macro- and micronutrient goals for the child. There are online and app-based recipe builders using the exchange system. Alternatively, customized recipes can be developed, for instance by using the Blenderized Diet Recipe Calculator (11).

Milk or infant formula can serve as a base for a blenderized diet, which can contain a variety of foods. A recipe should be made to guide food choices and portions to best meet nutritional needs. If the child is able to take oral feeds by spoon, the remainder can be supplemented via tube. Home-blended formulas are best delivered through a gastrostomy tube because these feedings are viscous and may clog a narrow nasogastric tube.

Table B-2: Easy Blenderized Tube Feeding Recipe

Ingredients	
• 3 jars (2.5 oz each) strained baby food meat • 3 jars (4 oz) baby vegetables – one should be green leafy green, e.g., spinach, and one orange, e.g., squash, pumpkin, sweet potato, carrots • 3 jars (4 oz) baby fruits • 2 tablespoons olive oil	• 2.5 cups whole milk • 6 tablespoons infant cereal (oatmeal, barley) • 7 tablespoons nonfat dry milk powder • 1 tablespoon dark corn syrup
Delivery	
Yield: 1500 cc or 50 oz **Energy:** 1500 kcal or 30 kcal/oz **Nutrients:** 66 grams protein (18% of calories) 67 grams fat (40% of calories) 159 grams carbohydrates (42% of calories) 10 grams fiber 100% of DRI for children ages 1-3 years.	

Note: A liquid vitamin or dissolved children's multiple vitamin/mineral supplement may be needed to meet the DRI's for older children, and a vitamin D supplement is needed for all children. It is best to use a variety of baby meats, vegetables, fruits, and grains to meet nutritional needs and mitigate consumption of heavy metals. Use of this recipe should be assessed by the child's physician and RDN.

Modular Products

Modular formulas or products are not nutritionally complete. They contain specific nutrients, which can be added to commercial or home-prepared formulas. Examples of modular products include the following:

- Fats: provide additional energy. Medium-chain triglycerides are fats that do not require bile acids and lipase for digestion and absorption. If the child can digest fats, a simple vegetable oil can be used.
- Readily-digested carbohydrates: provide additional energy
- Protein and specific amino acid preparations: increase protein intake.

Careful monitoring of modulars is important to ensure that all nutrient needs are being met and that the formula is tolerated with the additions. See Appendix F for examples of modular products.

Specialized Formulas

Specialized formulas are available for children and youth with specific needs, such as prematurity, renal failure, or inborn errors of metabolism. A physician or RDN who is familiar with the products and their particular uses should select the formula.

Additional formula thoughts

The more viscous a formula, the slower it will flow through the tube. This may make it more difficult to gravity feed/bolus feed the formula, so parents may add water to "thin" the formula and increase the rate of flow. Another alternative is to use an enteral pump for bolus feeds. Formula that is cold is also more viscous and may cause stomach distress.

When instructing parents on formula use, written recipes are very helpful, along with making sure parents have appropriate measuring utensils. For example, a soup spoon does not equal a teaspoon. Have parents demonstrate how they mix the formula. Often what is assumed is being delivered to the child is very different from what is actually delivered.

Cleanliness is very important in preventing bacterial contamination and formula tolerance. Multiple episodes of diarrhea may indicate bacterial contamination and not viral illness or formula intolerance. Hang times for sterile formula are 8-12 hours with aseptic handling. If you are using powdered formula, the hang time is 4 hours. Please check with the formula company regarding hang times. The Pediatric Nutrition Practice Group from the Academy of Nutrition and Dietetics published the third edition of Infant and Pediatric Feedings: Guidelines for Preparation of Human Milk and Formula and in Health Care Facilities in 2018 (7).

Administration of Feeding

Formula delivery will depend on tolerance, volume requirements/limits, safety, and the family's home schedule. What works for a hospital setting may not be appropriate for the child's home, school, therapy, and/or daycare schedule. Adherence often relies on simplifying enteral feeding for the family.

Children beginning tube feedings may be started on full strength isotonic formulas, given in small volumes (see recommended rate below). Hypertonic formulas should be started at half strength. Some children who have not had oral or tube feedings for a long period of time or have a history of formula intolerance (such as premature infants or children with short gut syndrome) may require half-strength formula initially, with gradual increases to full strength. In general, if a child needs diluted formula, it is best to increase the volume to make sure the child's fluid needs are met then gradually increase the concentration. Concentration and volume should not be increased at the same time. Frequent adjustments may be necessary as the child adjusts and as the family schedule changes.

Suggested schedule to initiate enteral feedings (4):

- Infants 10 ml/hour
- Child 1-5 years 20 ml/hour
- Child 5-10 years 30 ml/hour
- Child >10 years 50 ml/hour

Advance the delivery rate as tolerated to meet the goal for the child's nutrition needs. Increase volume every 4-12 hours, monitoring carefully for tolerance. Tolerance is defined as the absence of diarrhea, abdominal distension, vomiting, or gagging. If a child is bolus fed, start feeds at 25% of goal volume, increasing as tolerated.
Venting often improves tolerance to feeds. Venting is letting the air out of the stomach via the feeding tube, i.e., "burping" by attaching a 60 ml syringe and massaging the tummy. The Farrell Decompression Valve system enables the child to vent the stomach while receiving tube feedings.

The physician may require that residuals be checked when a tube feeding is initiated or when formula or medications are changed. Gastric residuals are primarily used to check placement for NG tubes (12,13). There is minimal information on the risks/benefits of routinely measuring gastric residual volume, so many are abandoning this practice.

Monitoring

Monitoring home enteral feeding for children varies greatly. Often, enteral supplies and formula are delivered to the home, which may be the only point of contact among a child, their family, and the home care provider. Children on enteral feeds are a high-risk group that requires closer monitoring. What works best for the child and family is a team of health care professionals who work together. In Washington State, nutrition assessment and monitoring is reimbursable for children with Medicaid services. However, for many children with private insurance, enteral formula or nutrition assessments may not be reimbursable benefits because enteral feeding is considered food.

Medications and Tube Feedings

The goal for children on enteral feeds is to optimize nutrition therapy and maximize therapeutic response to medications administered through the feeding tube. Children on enteral feedings often have extensive medication regimens; a benefit of tube feeding is delivery of medication by tube. The child does not have the opportunity to refuse to swallow, drool, or vomit medication, so they receive all of the prescribed medication with better efficacy. Feeding tubes may be a convenient avenue for medications, but some may be incompatible, causing the tube to clog, interacting negatively with specific nutrients, or affecting feeding tolerance. Formula or its components can also interfere with the absorption, distribution, metabolism, or elimination of medication (see Chapter 5). It is important for the pharmacist and physician to know that the child is receiving medication via their gastrostomy tube. The pharmacist and RDN can review the medication list for compatibility with enteral feeds.

Medications should not be added to enteral formulas. Avoid mixing medications together. Each medication should be administered separately. Families should be encouraged to give medicine in a liquid form. Elixirs and suspensions can usually be delivered through the feeding tube without

a problem. Compressed tablets can be crushed and mixed with water. Pill crushers are helpful, or you may let sit for 30+ minutes to dissolve. Syrups are incompatible with tube feedings because they tend to clog the tube unless diluted with water. Solid medicines, such as sustained-action tablets or capsules, or enteric-coated tablets, should not be crushed and delivered through the tube; once crushed, their action may be altered or they may cause gastrointestinal distress. Check with the child's physician for another medication preparation.

Medications are a common cause of tube occlusion. To maintain patency of feeding tubes, flush regularly with water, i.e., before, after, and in between giving medications. Another common side effect with medications is feeding intolerance or GI discomfort. Review medications to determine when it is best to administer each one. Adjustments to the feeding and medication schedules may be needed to improve tolerance and comfort. Table B-3 covers common tube feeding complications. For more information on medications and drug-nutrient interactions, see Chapter 5.

Daily Care of Tube

Contact health care provider regarding care instructions. See Table B-3 for some common complications of tube feeding.

Before feeding:

- Wash hands with soap and water before feeding
- Gather supplies needed for tube feeding; formula should be at room temperature
- Inspect site for skin irritation or leakage
- Check the tube for inward/outward migration
- Clean site with plain water or simple soap and water in circular motion away from stoma site
- Dry site
- If needed, stabilize tube with gauze and tape

Typical Gastrostomy Feeding

Position child with head higher than stomach, upright, or on their side. An infant seat, high chair, or pillow or wedge for propping may be helpful. Check residuals if recommended by the physician. Residuals may need to be checked with new tube feedings or when switching to a new formula. Residuals may also be checked if the child appears to have or complains of nausea or fullness before the next feeding. There is minimal information on the risks/benefits of routinely measuring gastric residual volume, so many are abandoning this practice (12,13).

Bolus feeding

A feeding should take 15-30 minutes. If given too quickly, the child may experience sweating, nausea, vomiting, or diarrhea.

- Syringe: Attach syringe to feeding tube, pour formula into syringe. You may need to push with plunger to start flow and fill tubing. You do not want air in tubing. Connect filled tubing to gastrostomy. Control rate of feeding by raising or lowering syringe. Continue adding formula to syringe until total feeding is given.
- Feeding Bag: Clamp tubing, fill bag and tubing with formula. You may need to squeeze bag to start feeding. Control rate of flow with clamp. Hang bag from IV pole.
- The feeding tube may be left in place, unclamped to allow the child to burp, after about 10-30 minutes. If the child has a button gastrostomy, he will need a decompression tube to vent air. To prevent reflux, the child may need to remain with head elevated 30-60 minutes after feeding.
- After formula and burping are finished, flush tubing with 10-30 cc water. Close tube. Tuck gastrostomy under clothing.
- Wash feeding set with hot, soapy water, rinse well, and air dry. Feeding sets may be reused.

Continuous drip

- Clamp tubing on feeding bag and fill with formula. Unclamp tubing and fill drip chamber 1/3 full, then fill remaining tubing with formula to minimize air into stomach and clamp. Thread tubing through pump. Connect to gastrostomy tube. Unclamp feeding tube and start pump. The home care supply company will have instructions on how to use the pump. Feedings should not hang for more than 4-8 hours. On hot days, ice can be slipped into the pocket of a feeding bag to keep formula cool.
- Children can be cuddled or held during feeding. Include the child at family mealtimes. To distract the child while feeding or doing skin care, play games or music, tell a story, offer toys, etc. Oral motor stimulation is recommended.
- You may want to secure tube connections with tape so they do not come apart. Securing tube to clothing, out of reach of the child is helpful. Tubing can be tucked under clothing (onesies, overalls, tube tops and bandnets are helpful).
- The feeding tube may be left in place, unclamped to allow the child to burp, after about 10-30 minutes. If the child has a button gastrostomy, he will need a decompression tube to vent air. To prevent reflux, the child may need to remain with head elevated 30- 60 minutes after feeding.
- After formula and burping are finished, flush tubing with 10-30 cc water. Close tube. Tuck gastrostomy under clothing.
- Wash feeding set with hot, soapy water, rinse well, and air dry. Feeding sets may be reused.

Skin/Stoma Care

To prevent irritation/infections, the medical team will have instructions for new tube placement and site care. Once a stoma site has healed, a bath with warm soapy water may be all that is needed.

When to Call the Doctor

- If the skin around the gastrostomy is warm, tender, bright red – larger than a quarter
- If excess puffy red tissue is building up around stoma site or persistent bleeding around stoma site
- If there is excess leaking around stoma site or tube, e.g., soaking 2x2 gauze in <4 hours
- If stomach contents are leaking through button
- If child has persistent vomiting, diarrhea, or constipation
- If the feeding tube is blocked and you cannot remove blockage
- If the feeding tube is pulled out
- If the child has a temperature >101˚F

Common Pump Problems—Check this list if the pump isn't working correctly

- Did the "START" button get pressed?
- Are the clamps open?
- Is the tubing kinked?
- Is the drip chamber too full or not positioned correctly?
- Is the "Pause" button on?
- Is the feeding tube plugged?
- If none of the above, call home care agency.

Oral Hygiene

Oral hygiene is important even if the child is not eating by mouth. Proper oral hygiene cares for the teeth and prevents cavities; it is also an important part of oral motor stimulation for the child.

Social Concerns with Tube Feeding

An important consideration in tube feeding is the family's ability and willingness to carry out the tube feeding program. Concerns include the availability and cost of equipment and formula, home sanitation and family hygiene, family support

systems, and other psychosocial factors. Many families have a difficult time deciding to use a tube

for feeding their child. When families are asked about tube feeding, their concerns include finding a caregiver to tube feed their child, public ignorance about tube feeding, planning their social life around feeding schedules, and
sadness over depriving their child of the pleasure of eating by mouth. Reviewing the benefits of tube feeding and allowing them to talk with other parents may help decrease their anxiety. Insurance coverage for formulas and feeding equipment should be determined before the child is hospitalized for tube placement.

Before the child is discharged from the hospital, the caregiver(s) must be prepared for tube feeding. More than one family member or caregiver should be taught about the tube feeding to ensure continuity of the child's feeding program and to prevent isolation of the primary caregiver. Caregivers should be thoroughly instructed on the following aspects of tube feeding: formula preparation, use and care of equipment, insertion of the tube, stoma care, and emergency procedures. The caregivers should be encouraged to keep the following records in a notebook, which they should bring to each clinic visit: formula intake, stooling pattern, activity, behavior, medications, and instructions from medical staff. Identify who will provide formula, supplies, and nutrition follow-up. A home care company can provide feeding supplies and equipment. The Special Supplemental Nutrition Program for Women, Infants and Children (WIC) may provide some formulas to eligible infants and children.

The caregiver(s) should be contacted daily for the first week the child is home, or until they feel secure with the tube feeding regimen. Follow-up can be provided by home visit, clinic visit, or telephone. The caregiver(s) should be given a phone number for 24-hour assistance regarding problems with tube feeding.

Family meals offer important learning experiences for children who are tube fed. It is important for the child to associate the satisfying feeling of fullness with the pleasant time of family meals, including social interactions, good smells, and the appearance of food. Even if the child does not experience the tastes and textures of oral feeding, social experiences can be provided. This is important if the child is to eventually transition from enteral to oral feeding.

Feeding Behaviors

Negative or atypical feeding behaviors may be present before a child is tube fed and additional behaviors may develop while the child is tube fed. See Chapter 9.

School and Tube Feeding

Children with feeding tubes are eligible for expanded nutrition services in schools through Public Law 99-457 and the Americans with Disabilities Act. Tube feedings can be given as a routine activity at school. This presents understandable concern for educators. A team, including an RDN,

special educator, nurse and the family can help facilitate feeding in the school. The objective is to use the same feeding routine, positioning, and oral-motor stimulation at home and at school. The physician's orders, an individualized education plan (IEP), and instruction on when to call the family or physician may be required. See Chapter 12.

Transition to Oral Feeding

Transition to eating by mouth starts when the tube is first placed. All children on tube feedings require oral stimulation for development of feeding skills (see Chapter 8). The child may need to "re-learn" that food in his mouth can satisfy hunger, plus keep his mouth "awake" and ready to accept foods. Transition is generally most successful when the process involves a team; a team might include a pediatrician, RDN, feeding therapist, and nurse. Successful treatment addresses the following questions:

- Can the child eat safely? How are their oral skills?
- Has the child shown appropriate growth on enteral feedings? Often, a child will not show hunger until an appropriate weight for height is reached.
- Has the medical condition for which the child had a tube placed resolved or improved?
- Are the parent and child ready to transition? Do they have the time to devote to transitioning?

A common approach to transition is to begin by promoting the child's recognition of hunger cues. If necessary, "normalize" the tube feeding schedule to include three large and two to three small bolus feedings each day, resembling meals and snacks. This will help the child to recognize hunger and satiety. To stimulate hunger, caloric intake may need to be decreased by 25% and/or night-time feeds, discontinued. Oral intake may be minimal, so supplemental feedings are offered via the tube after each meal. As the child is able to consume more food orally, the tube feeding can be decreased. It is important to ensure an adequate fluid intake—continue to meet water needs by tube or orally.

It takes time to change feeding behaviors. The longer a child goes without eating by mouth, the longer it will take to transition to oral feeding. Small steps are important, so the child feels that they are in control. The child has the benefit of using the feeding tube to meet their nutritional requirements (9).

The tube can be removed when the child can eat an adequate amount of food orally to support growth. It may be prudent to wait until the child shows that they do not lose excessive weight with illness. Weaning from tube feeding is often a time when a child may not be meeting nutrition or hydration needs and will require increased monitoring. When a child demonstrates oral eating to support growth, their medical team may remove the feeding tube. A gastrostomy tube may be simply pulled, and the stoma will gradually close on its own. The skin can be protected from leaking with gauze and barrier creams. If the gastrostomy stoma does not close on its own, the

stoma may need to be closed surgically. This may happen if the child has had a g-tube for a long period of time or other stoma site problems. Jejunostomy stomas will need to be closed surgically.

Table B-3: Common Complications of Tube Feeding

Complication	Possible Cause	Intervention
Nausea/Vomiting and Diarrhea	Rapid administration of feeding	• For continuous drip feeding, return infusion rate to previous tolerated level, then gradually increase rate • For bolus feeding, increase length of time for feeding; allow short break during feeding; offer smaller and more frequent feedings
	Hyperosmolar solution (energy-dense and/or high protein formulas)	• Switch to isotonic formula • Dilute current formula to isotonic, and gradually increase to full strength • Check that formula is mixed properly • Avoid adding other foods to formula, i.e. baby food, powdered formula
	Medication	• Do not add medication to formula; give between feeding with water or juice • Medications that may cause diarrhea include antibiotics, GI neurologic stimulants, beta blockers, stool softeners, liquid medications with sorbitol • Review medication profile and make recommendations for changes
	Air in stomach/ intestine	• Burp child during feedings or allow for short breaks • Use medication to decrease gas, i.e., simethicone • Elevate child's head during feeding and for 30 minutes after meal
	Tube migration from stomach to small intestine	• Pull on tube to reposition against stomach wall
	Cold formula	• Warm formula to room temperature
	Rapid GI transit	• Select fiber enriched formula
	Bacterial contamination	• Refrigerate open cans of formula, keep only as long as manufacturer suggests • Clean tops of formula cans before opening • Hang only a 4 hour amount of formula at a time • Be sure feeding sets are cleaned well
	Allergy/lactose intolerance	• Try a lactose-free formula • Try soy formula; if allergic to soy, try elemental or semi-elemental formula
	Excessive flavorings	• Stop using flavorings
	Excessive fat	• Decrease fat in formula or use MCT oil • Refer to physician
Constipation*	Inadequate fiber/bulk or fluid	• Try formula with added fiber • Increase water • Supplement with prune juice • Try stool softeners, suppositories, or enema, as indicated • Refer to physician

* See Chapter 6.

continued...

Complication (continued)	Possible Cause	Intervention
Gastroesophageal reflux	Delayed gastric emptying	• Refer to physician • Recommend medication to stimulate movement of GI tract • Elevate child's head (30-45°) during feeding and for 1 hour after meal • Check for residuals before feeding • Try smaller, more frequent bolus feedings or continuous drip feeding • Consider jejunal feeding
Large residuals	Decreased gastric motility	• Elevate child's head during feeding • Use gastric stimulant to promote gastric emptying • Consider continuous feeds
	Hyperosmolar formula	• Switch to isotonic formula
	Medications	• Do not add medications to formula; give between feeding with water or juice. • Refer to physician
Tube feeding syndrome (dehydration, azotemia, and hypernatremia)	Excessive protein intake with inadequate fluid intake	• Refer to physician • Decrease protein • Increase fluids. Monitor fluid intake and output
Hyponatremia	Inadequate serum sodium may be related to excessive water intake, excessive fluid loss (e.g., vomiting and/or diarrhea), medications, tracheostomies, or fluid retention related to renal or cardiac problems	• Refer to physician • Replace sodium losses • Restrict fluids
Clogged feeding tube	• Formula residue or coagulated protein • Inadequate flushing of tube • Medication	• Use correct formula • Flush tubes with water after giving formula or medication • Flush every 3-4 hours with continuous drip feeds • Do not mix formula with medication • Irrigate with air, using syringe • Gently "milk" tubing • Dissolve ¼ tsp. meat tenderizer in 10 cc water and flush to dissolve clot • Replace tube
Leakage of gastric contents	• Improper positioning • Tube migration • Increased size of stoma	• Place child upright for feeding • Make sure gastrostomy tube is firmly in place • Stabilize tube with gauze pads, adjust crosspiece • If stoma is too large for tube, insert new tube • Keep skin around stoma clean and dry; use protective ointments and gauze • If leaking out of button gastrostomy, may need to replace device • Refer to physician
Bleeding around stoma	• Excessive movement or pressure on tubing	• A small amount of bleeding is normal • Tape tube securely in place to avoid irritation from movement • Secure tube under child's clothing • Refer to physician

Complication	Possible Cause	Intervention
Infection of stoma	• Gastric leakage around tube • Stoma site not kept clean • Allergic reaction to soap	• Correct cause of leakage • Carefully cleanse and protect stoma • If stoma site is irritated, use plain water or change soap used • Refer to physician for culture and medication
Granulation tissue	• Body rejecting foreign body • Poorly fitting tube causing friction • Use of antiseizure medication, such as Dilantin	• Keep area clean and dry • Adjust snugness of PEG tube with crosspiece • Stabilize tube using tape, bandnet, ace bandage, tube top • Prevent child from pulling on tube • Apply silver nitrate as directed by physician
Bad Breath	• Poor oral hygiene • Dry mouth, dehydration • Bacterial overgrowth • Infection	• Brush and floss daily • Reassess fluid needs • Contact doctor

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Resources

ASPEN Enteral Nutrition Resources
https://www.nutritioncare.org/Guidelines_and_Clinical_Resources/Enteral_Nutrition_Resources/
This site has instructional videos for Enteral tube placement in English/Spanish and Chinese. Videos are available for professionals and for caregivers. There are videos on how to make blended tube feedings, how to use different pumps, give medications by tube, and more.

Basics of Homemade Blenderized Tube feeding Preparations Video
https://www.youtube.com/watch?v=KhZfgaCtJVo
This video discusses creating homemade blenderized tube feeding (BTF), food-borne infection considerations, and food safety and sanitation procedures.

Appendix C

TECHNICAL ASPECTS OF HOME PARENTERAL NUTRITION

Kathryn L. Hunt, RDN, CD

The American Society for Parenteral and Enteral Nutrition (ASPEN) defines parenteral nutrition (PN) as nutrients provided intravenously. The most commonly used solutions are a combination of dextrose (carbohydrate), amino acids (protein), and fat (lipids). Parenteral nutrition can refer to all three components or the components dextrose and amino acids only. This section describes methods used to determine the components of a parenteral nutrition solution.

Fluid

The initial step in determining the PN solution is the estimation of the patient's fluid needs. General guidelines for fluid management of PN in older infants and children are outlined in Table C-1 (1).

Infants on PN often have increased energy needs due to the natural demands of growth combined with the stress of illness. PN solutions providing "maintenance" levels of fluid may not meet such energy needs and may be too hypertonic. For example, infants with illness or under stress often need between 135-150 mL/kg/day of total fluid to adequately meet their energy needs for growth. Therefore, fluid volumes may be administered in excess of maintenance calculations and then macronutrients may be increased until the solution reaches the desired energy goal.

Table C-1: Estimating Fluid Requirements: Older infant/child

Child's weight (kg)	Fluid Requirements Per Day
<10 kg	100 ml/kg/day (infants may need up to 135-150 mL/kg/d)
10-20 kg	1000 ml + (50 ml/kg for every kg between 10-20 kg)
>20 kg	1500 ml + (20 ml/kg for every kg > 20)
>40 kg	1500 ml per m2 body surface area

Some conditions increase fluids needs:

- Fever (any degree of fever above normal (37.0-37.4 C) needs immediate medical attention. Fevers are often indicators of line sepsis.)

- Hypermetabolism
- GI losses (diarrhea, ostomies, vomiting)

Some conditions decrease fluid needs:

- Heart disease
- Renal failure with low urine output
- Hepatorenal syndrome

Energy

Parenteral energy needs vary depending upon the activity and stress of the individual child. Because PN requires less energy for digestion and absorption, energy needs in stable (renourished and not stressed), parenterally fed children may be up to 15% lower than for those who are enterally fed. Conversely, an individual child's energy needs may be higher than the DRIs during periods of catch-up growth, hypermetabolism, and illness (2,3).

The dietitian must monitor the child's rate of weight gain to ensure appropriate growth. Only by tracking monthly plots on the child's growth chart can the dietitian determine whether to increase energy to maintain the growth pattern or decrease calories to prevent excessive weight gain.

Other Nutrients

Protein in the form of **amino acids (AA)** provides 4 kcal/g and should provide 6-16% of total energy, depending upon the child's energy needs and disease state. For infants and children, AA are typically started at 1.5 g/kg/day and increased by increments of 0.5 g/kg/d until the final protein goal is reached (2).

Fat in the form of intravenous lipids **(IV)** provides a concentrated, iso-osmolar source of energy, supplying approximately 20-40% of the child's total calories. In addition to calories, fats provide essential fatty acids, i.e., linoleic acid (LA, omega-6) and α-linolenic acid (ALA, omega-3), both of which cannot be synthesized in the body and are required in the diet. The minimum amount of lipid required to prevent essential fatty acid deficiency is 4-7% of total calories, while the maximum amount of lipid that may be administered is 60% of total calories (4). Soybean oil with high omega-6 fatty acid content is an excellent source of these essential fatty acids and has long been the standard formulation used in most patients; however, newer options are now available with more balanced fatty acid profiles (4). Such IV lipids are available as 20% emulsions and provide 2 kcal/ml.

Carbohydrate (dextrose) is typically the major source of non-protein energy and provides 3.4 kcal/g. In general, dextrose should provide 40-50% of total energy (3).

Electrolytes and minerals are adjusted in PN solutions based on serum lab values. The team managing the child's PN solution will determine the amounts of electrolytes and minerals to be included in the solution.

Vitamins are added to each bag of PN solution prior to administration. The PN solution is clear until the vitamins are added; the vitamins change the color of the PN solution to yellow. Patients on PN support must receive adequate vitamin supplementation, and current parenteral multivitamin products commercially available in the U.S. meet the requirements for most PN patients. An exception may be patients who require more than the standard daily dosing of vitamin D to correct deficiency. Currently no separate parenteral vitamin D products have been developed for PN-dependent patients who fail to respond to oral vitamin D therapy (6). Recommended parenteral vitamin and mineral intake levels are provided in Tables C-2 and C-3 (5).

Trace elements included in the PN solution will vary, depending on the child's age, size, and medical condition. For example, patients with large stool and ostomy outputs may require additional zinc. In general, children are not discharged from the hospital until their medical conditions are stable and PN solutions are firmly established. Recommended trace element intake levels are provided in Table C-4 (5).

Table C-2: Recommended Parenteral Vitamin Intakes for Term Infants and Children (6)

Vitamin Requirements		
	Less than 11 years old MVI Pediatric Dosing: 2 mL/kg (Max 5 mL/day)	≥11 years old: MVI-Adult (Multi-12) Dosing 10 mL/day
Vitamin	**Content per 5 mL**	**Content per 10 mL**
Vitamin A (IU/d)	2300	3300
Vitamin E (IU/d)	7	10
Vitamin K (phytonadione)(µg/d)	200	150
Vitamin D (IU/d)	400	200
Ascorbic acid (mg/d)	80	200
B1 (Thiamin) (mg/d)	1.2	6
B2 (Riboflavin) (mg/d)	1.4	3.6
B6 (Pyridoxine) (mg/d)	1	6
B3 (Niacin) (mg/d)	17	40
Pantothenic Acid(mg/d)	5	15
Biotin (µg/d)	20	60
Folic acid (µg/d)	140	600
B12 (Cyanocobalamin) (µg/d)	1	5

Table C-3: Recommended Parenteral Mineral Intakes for Term Infants and Children (5, 6)

Nutrient	Term Infant (<1 yr)	Children (>1 yr)	Adolescents
Calcium (mEq/kg/d)	0.5-4	0.5-4	10-20
Phosphorus (mMol/kg/d)*	0.5-2	0.5-2	10-40
Magnesium (mEq/kg/d)	0.3-0.5	0.3-0.5	10-30

*Phosphate requirements are always represented as millimoles of phosphate:

Potassium Phosphate	Sodium Phosphate
1 mL = 3 mMoL phosphate = 93 mg elemental phosphorus 1 mL = 4.4 mEq potassium = 170 mg potassium	1 mL = 3 mMol phosphate = 93 mg elemental phosphorus 1 mL = 4 mEq sodium = 92 mg elemental sodium

Table C-4: Recommended Parenteral Trace Element Intakes for Term Infants and Children

Adapted from ASPEN 2012 Trace Element Guidelines: Normal Requirements (6)

Trace Element	Infants ≤ 2 kgs	Infants >2 kgs-6 months	Infants 7-12 months	1-3 years	4-8 years	9-13 years	14-18 years
Chromium (mcg/day)	None	None	None	0.3	0.3	0.5	0.5
Copper (mcg/kg)	20	20	20	5-20	5-20	200-500 mcg/day	300-500 mcg/day
Manganese (mcg/kg)	0.5-1	0.5-1	0.5-1	0.5-1	1 Max of 55 mcg/day	1 Max of 55 mcg/day	55 mcg/day
Zinc (mcg/kg)	400	250-400	250	125	125 Max of 5 mg/day	5 mg/day	5 mg/day
Selenium (mcg/kg)	3	2	2	2	2	60-100 mcg/day	60-100 mcg/day

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Appendix D

DIET ORDER FOR MEALS AT SCHOOL

Student's name_____Age_____Grade_____
Disability_____
Other Medical conditions_____

Diet Order (check all that apply):

- ☐ Increased calorie #kcal
- ☐ Decreased calorie #kcal
- ☐ PKU
- ☐ Food allergy
- ☐ Other:_____

Texture Modification

- ☐ Soft and bite sized
- ☐ Minced and Moist
- ☐ Pureed

- ☐ Thickened Liquids
- ☐ Tube feeding
  - ☐ Liquified Meal
  - ☐ Formula_____type

Foods to Omit	Foods to Substitute

I certify that the above-named student needs special school meals prepared as described above because of the student's disability or chronic medical condition.

_____Physician or recognized medical authority signature (circle)

Office phone number_____Date_____

Diet Prescription for Meals at School

Section 504 of the Rehabilitation Act of 1973 assures handicapped students access to school meal service, even if special meals are needed because of their handicap. If special meals are needed and requested, certification from a medical doctor must:

1. Verify that special meals are needed because of a disability or medical condition
2. Prescribe the modified diet and/or textures allowed

Name of student for whom special meals at school are requested:

_____

Disability or medical condition that requires the student to have a special diet:

_____

_____

Foods Prescribed: _____

_____

_____

Texture Consistency Required (note solid and liquid consistency requirement):

_____

_____

Feeding Positioning and Assistance:

_____

_____

_____

Other Information Regarding Diet or Feeding (Please provide additional information on the back of this form or attach to this form.)

I certify that the above named student requires special school meals prepared as described above because of the student's disability or chronic medical condition.

_____	_____	_____
Physician/Recognized Medical Authority Signature	Office Phone Number	Date

Appendix E

IEP NUTRITION RELATED GOALS AND OBJECTIVES

Goals	Considerations for Writing Objectives
1. To develop or refine self-feeding skills	
• finger feeding • use of feeding utensils • use of a cup or glass	• identify special feeding equipment and level of assistance needed for practicing skills
2. To improve oral-motor function related to eating	
• lip closure: on spoon/fork on cup/glass while chewing at rest • tongue movement (within and outside mouth) lateralization, elevation and depression • chewing pattern • suck through straw • bite off piece of food • oral reflexes, hyperactive gag, tongue thrust • oral sensitivity hyposensitive hypersensitive	• consider exercises to facilitate oral-motor function and specify frequency and duration of the exercises • consider food texture and consistency changes to facilitate improved oral-motor function • identify positive reinforcement for successful attempts
3. To improve mealtime behaviors	
• inappropriate finger feeding • pace • rumination/regurgitation • food acceptance (textures, types or variety of foods) • neatness • feeding posture/position • self-abusive behavior • staying on task • staying at the table	• identify supervision needed for monitoring mealtime behavior • identify presentation of new foods/textures and situations • identify the appropriate mealtime environment • identify positive reinforcement for appropriate behavior
4. To identify and communicate nutrition needs	
• hunger • thirst • food names • food groups • restricted foods • special nutrients (iron rich foods, etc.)	• identify special instruction or learning activities to teach nutritional needs • identify games/exercises for food or nutrient recognition • identify positive reinforcement for correct responses
5. To improve food preparation and meal time skills	
• open can, box, carton package • make sandwich • make snack • pour, stir, slice, etc. • set table • clear table • clean table, utensils	• identify practice periods and exercises or steps for skill development • identify supervision needed to monitor skill development • identify positive reinforcement for successful attempts

continued...

6. To improve growth rates	
• weight maintenance with continued linear growth • gradual weight loss with continued linear growth • weight gain and linear growth	• identify person(s) responsible for tracking growth • develop a school weight control program • identify dietary supplements and modifications provided by the school and/or the family • identify extra snacks scheduled during the school day • identify positive reinforcers for growth changes
7. To maintain lab data within normal limits*	
• blood glucose levels (Diabetes) • phenylalanine levels (PKU)	• identify a method to obtain and communicate specific lab values • identify methods of monitoring this data

*These are very specific nutrition goals which may not be appropriate for the school to monitor. However, other goals may be written which relate to this data.
Example: For the child with diabetes, the goal is to decrease episodes of hypoglycemia.

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Appendix F

COMMERCIAL NUTRITION PRODUCTS

Kim Nowak-Cooperman, MS, RD, CD and Sarah Bailey Harsh, MS, RDN, CD

This section contains general information about commercial nutrition products. Manufacturing processes and products change frequently and a list of specific products is beyond the scope of this publication. The resource section includes lists of commercial nutrition products maintained by other organizations.

Infant Formula

Human Milk- Human milk is safe for human infants except for in the few circumstances outlined by the CDC in their document "Contraindication to Breastfeeding or Feeding Expressed Breast Milk to Infants." The complete document is linked in the references (1).

Standard Milk- Based Infant Formulas- Indicated for most infants. Standard milk-based formulas generally contain whey and casein, medium and long-chain fatty acids, and lactose.

Soy-Based Infant Formulas: Indicated for infants with galactosemia or lactase deficiency. Both are rare in infants. Soy-based formulas contain soy protein isolate, long chain fatty acids, and sucrose or glucose polymers. Soy infant formulas are contraindicated in preterm infants because of an increased risk of osteopenia. Infants with a milk allergy or cow milk protein induced enteropathy/enterocolitis may also react to soy protein (2).

Partially Hydrolyzed Formulas: The whey or casein-based proteins in these formula are hydrolyzed, or broken into smaller pieces. The smaller pieces may be easier for some infants to digest. These formulas are not appropriate for infants with cow's milk allergies.

Extensively Hydrolyzed or Amino Acid Based Formulas: The protein in these formulas is broken down extensively or is comprised of individual amino acids. These formulas are considered hypoallergenic and can be used for infants with cow's milk allergy.

Specialized Formulas: These formulas are modified for infants with specific electrolyte or macronutrient needs different than those of most infants. These made include those with kidney or heart disease, those on a specific diet for an inborn error of metabolism, or those being treated with a ketogenic diet.

Post Discharge Formulas for Premature Infants: These formulas provide higher calorie, protein and mineral content.

Follow-up Formulas (Toddler Formulas): These formulas are alternatives to cow's milk or soy milk. There is no indication for the use of these formulas in most children. Most are not nutritionally complete and they are not regulated the same way that infant formula is regulated (3).

Pediatric Formula

Standard Pediatric Formulas- Are nutritionally complete or meet the DRI for a given age group for all vitamins and minerals if a stated volume is consumed. Some products are flavored and some are unflavored for enteral use. A growing number of formulas have food-based ingredients.

Semi Elemental Formulas- The whey or casein-based proteins in these formulas are hydrolyzed or broken into smaller pieces that may be easier to digest.

Elemental Formulas- The protein in these formulas is comprised of individual amino acids. These formulas are considered hypoallergenic.

Specialized Products- These formulas are modified for children with specific electrolyte or macronutrient needs. These made include those with kidney or heart disease, those on a specific diet for an inborn error of metabolism, or those being treated with a ketogenic diet.

Modulars

These products are not nutritionally complete and are used to add extra fat, calories, protein or fiber to a formula or food.

Resources

Formula tables

1. ASPEN Infant Formula Guide- https://www.nutritioncare.org/Guidelines_and_Clinical_Resources/EN_Formula_Guide/EN_Infant_Formulas/

2. ASPEN Pediatric Formula Guide - https://www.nutritioncare.org/Guidelines_and_Clinical_Resources/EN_Formula_Guide/EN_Pediatric_Formulas/

3. ASPEN Modulars guide- https://www.nutritioncare.org/Guidelines_and_Clinical_Resources/EN_Formula_Guide/EN_Modular_Products/

4. AND's Pediatric Nutrition Practice Group (PNPG) Infant formula tables: Includes nutrients, measurements and recipe tools- (https://www.pnpg.org/pnpg/resources/infant-formula-tables)

Formula Manufacturers

Abbott Laboratories, Ross Products Division:
Product information at http://abbottnutrition.com/
1-800- 227-5767; M - F, 8:30 a.m. to 5 p.m. EST:
Columbus, OH

Earth's Best:
Product information at www.earthsbest.com

Functional Formularies:
Product information at www.functionalformularies.com

Happy Baby:
Product information at www.happyfamilyorganics.com

HiPP:
Product information at www.hipp.com

Holle:
Product information at https://gb.holle.ch

Hormel Health Labs:
Product information at http://hormelhealthlabs.com
Savannah, GA
1-800-866-7757

Loulouka:
Product information at https://loulouka.nl

Kate Farms:
Product information at www.katefarms.com

Kendamil:
Product information at www.kendamil.com

Mead Johnson Nutritionals:
Product information at http://www.meadjohnson.com
Phone: 812/429-5000; M-F 7:30-4:00, Central Time
Toll Free: 800/BABY-123

Nestle Nutrition:
Product information at http://nestlenutrition.com/us
Toll Free: 800/422-ASK2 (2752)
Minnetonka, MN

Nutricia International, North America:
Product information at http://www.nutricia-na.com
Toll Free: 800-365-7354
Gaithersburg, MD

Precision Foods (Thick It and Thick It 2):
Toll Free: 800/442-5242 x258
Product information at http://www.thickitretail.com
St Louis, MO

Simply Thick:
Product information at simplythick.com

Vitaflo USA:
Product information at http://www.vitaflousa.com
1-888-VITAFLO
Huntington, NY

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