Children and Youth with Complex Cerebral Palsy

Care and Management





Edited by Laurie J. Glader and Richard D. Stevenson

A practical guide from Mac Keith Press

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From a Parent

When my triplets were born, three teams of medical professionals whisked them away before I could even lay eyes on them. And so in a moment, I had to learn to trust others with the well-being of my children. I had no idea how necessary this lesson would prove in the life of our family. When the boys were nine months old, I asked their occupational therapist why she was working so hard to get them to grab beads rather than working on sitting up – a skill I desperately wanted them to learn. I was crying as I asked, petrified of what her answer would be. I will never forget how, with tears streaming down her own cheeks, she gently told me they had to reach across midline before we could hope for them to reach any further milestones. Her tears were a gift to me that day because I needed to know she cared for my children with a passion and that she valued being on our team. For 21 years, we have had nurses, doctors, therapists and surgeons at birthday parties, theatre performances, and even high school graduations because these professionals, who have so expertly cared for our children, are on our family team, in our circle of supporters. They are our hand-holders and cheerleaders. We are ever grateful.

Yes, you are on the frontlines of your patients' medical care. But more importantly, to the parents of a child with complex cerebral palsy, you are on the frontlines of hope.

Carole Shrader

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Feeding and nutrition

Mary C. Bickley, Eva Delaney and Valentina Intagliata

Introduction

Feeding disorder from oral-motor impairment is one of the comorbid impairments in children with cerebral palsy (CP). Prevalence varies among studies, but one estimate is that as many as 90% of preschool children with CP have oral-motor dysfunction (Reilly et al. 1996). Some studies report that even children with mild CP show evidence of oral-motor dysfunction impacting functional feeding skills (Gisel et al. 2000).

Children with CP have a spectrum of feeding abilities. Some children with oral-motor impairment are prohibitively unsafe to eat orally due to the risk of aspiration. Others cannot handle solids, but may be able to eat safely and grow adequately on purees or specific food/liquid consistencies. Many children with severe CP are inefficient eaters. These children may be able to chew and swallow safely, but may take excessive time to eat, and fatigue easily doing so.

In the most severe cases of feeding disorder, children require placement of a feeding tube to maintain adequate nutrition and/or to ensure safe feeding. Other children may be able to learn strategies to help them with certain problematic textures of food. Often, the stress of the feeding disorder can have a profound impact on the family dynamics around mealtimes and the child's feeding behaviors.

In this chapter, we provide an overview of feeding, nutrition and growth in children with complex CP including oral motor dysfunction, feeding assessment and interventions, evaluation of nutrition and physical growth. Other issues related to feeding and

nutrition are covered in Chapter 8 (dysphagia, constipation, gastroesophageal reflux, gastrostomy, fundoplication), Chapter 9 (drooling, aspiration) and Chapter 20 (decision making for gastrostomy placement).

Oral-motor impairment

Oral-motor impairment or dysfunction is defined by differences in strength, tone and sensation of the oro-pharynx that affect the coordination of motor movements for both speech and eating. It is associated with disorders of swallowing, known as dysphagia. The terms dysphagia and oral-motor dysfunction are frequently used interchangeably but should be differentiated. Both can exist independently of one another but quite often have a reciprocal relationship. Oral-motor dysfunction can result in the mistiming of the swallow leading to aspiration. Dysphagia can exist with or without oral-motor dysfunction, and is sometimes secondary to structural or postural elements that impact the oral, pharyngeal or esophageal phase of the swallow.

Oral-motor impairment often impacts on the efficiency and pleasure of eating, as well as the social dynamics of feeding. Eating is the physical act of biting, chewing and swallowing food, whereas feeding describes eating in the social context. These social factors include caregiver–child relationships as well as the mealtime environment. Feeding is one of the most important initial interactions between a caregiver and a child, and any disruption in this process can have a negative impact on the child and the entire family.

Maturation of oral-motor skills follows an organized hierarchy of steps. This process begins with a reflexive pattern for suckling during early infancy, and proceeds through a series of learned behaviors that result in the ability to bite, lateralize, chew and safely swallow foods. Each step requires practice in a supportive environment so that the child can master efficient, effective and pleasurable eating. Motor and sensory impairments at any stage can affect the process of learning to eat.

The abnormalities in tone and the difficulties with motor coordination common in children with CP negatively impact eating skills. In early infancy, oral-motor dysfunction may be a presenting feature of CP since these children have impairments in coordinating and sustaining the rhythmic sucking pattern needed for feeding. In these young infants, frequent choking is common as well as prolonged feeding time and decreased intake, often leading to poor growth. Some children may have minimal oral-motor dysfunction as young infants (with reflexive suckling), but later struggle when they fail to master the complex voluntary, learned motor movements necessary for safe and efficient biting, chewing and swallowing.

The presence of increased muscle tone can make graded control of mouth position difficult. In effect, opening of the mouth for food entry and chewing is impaired.

Lingual lateralization, which is essential for maneuvering higher textured foods to the molars for proper mastication, is often impaired. In the absence of appropriate lingual manipulation, the child may not be able to transition safely and efficiently to a variety of textures.

Children with hypotonia often have difficulty achieving a closed mouth posture and this alone can make eating difficult. Weakness and decreased tone can result in a sluggish suck/swallow/breathe pattern (reflexive suckling) often associated with choking and a weak cough. Silent aspiration is also a potential problem due to weakness and poor sensory awareness. These difficulties lead to inefficient, prolonged and often unsafe eating. Children who are mostly hypotonic demonstrate significantly slower transition to purees and table foods, but generally progress further in oral-motor skills than children with significant hypertonia.

Assessment of eating

The three stages of eating – oral, pharyngeal and esophageal – are important to consider when assessing oral-motor dysfunction and dysphagia. Box 7.1 shows the American Speech and Hearing Association (ASHA) description of potential difficulties in each stage of eating that may be present in children with CP.

Box 7.1 Potential difficulties in each stage of eating that may be present in children with cerebral palsy

- Oral stage
  - Difficulties with motor coordination for graded movements of the lips, jaw and tongue.
  - Differences in tone impacting efficient motor and or sensory processing of age-appropriate textures and foods.
  - Difficulties with sensory tolerance of tastes, textures and temperatures.
- Pharyngeal stage
  - Difficulties with safe and efficient passage of food/liquids past the airway.
  - Penetration of liquid or food into airway.
  - Aspiration of liquid or food into airway.
  - Pooling or coating of residue.
- Esophageal phase
  - Difficulties with efficient and effective passage of food or liquids to the stomach.
  - Slow transit of food/liquid through esophagus.
  - Reflux of food/liquid back into airway.
  - Spasm or inability of sphincters to open/close appropriately.

Children with CP most commonly experience challenges in the oral stage. Abnormal motor movements and postural alignment of the head, neck and trunk significantly impact presentation of food to the child and their ability to keep food contained in the mouth. The loss of foods frontally can result in inefficient oral intake and unsafe swallow. These difficulties are only optimized by proper positioning and hands-on techniques, and cannot be 'fixed' or cured. Other difficulties during the oral stage include wide jaw excursion, tongue protrusion/thrusting and impaired lingual lateralization. As a result, these children have poor control of the food bolus as they attempt to swallow and this often leads to aspiration.

Assessment of feeding should be completed by a speech-language therapist along with a physical and/or occupational therapist to maximize feeding position. Additional consultation with the nutritionist allows for optimization of the child's diet within the constraints of his/her oral-motor dysfunction. An oral-functional evaluation by the speech-language therapist provides information regarding food texture that is most efficient and safe for a child with oral-motor dysfunction. This evaluation can also determine which utensils might optimize the child's oral-motor potential.

During the oral-functional evaluation, the clinician should be cognizant of potential aspiration. Clinical signs of aspiration include coughing, cessation of breathing, watery eyes, wet/or gurgly vocal quality and oral mucosal color changes. Cervical auscultation is another method that is useful for identifying more subtle signs of aspiration. This method entails the placement of a stethoscope on the median line of the cricoid cartilage and the listener interprets swallowing sounds. Consensus supports cervical auscultation as a good tool to determine if video-fluoroscopic study (VFSS) is necessary (Leslie et al. 2007).

VFSS study is warranted when there are clinical signs of aspiration as listed above, or if, in the absence of clinical signs, a child repeatedly has pneumonia or respiratory issues or a history of structural issues. VFSS is also a useful evaluation tool for the pharyngeal and esophageal stages of swallow that cannot be observed clinically. VFSS is not indicated when a child has 'never' eaten in the past, when there is the complaint of frequent gagging or as a precursor to 'feeding therapy'. These issues can be addressed through a clinical feeding evaluation.

Classification of eating and drinking abilities

The Eating and Drinking Ability Classification System (EDACS) has been developed specifically for children with CP in order to provide a meaningful framework for distinguishing varying eating abilities in everyday life. The EDACS classifies an individual's usual performance rather maximal abilities in ideal circumstances. The classification scheme characterizes eating and drinking abilities at five different levels for children

with CP aged 3 years and older. The most useful aspect of this scale is that it correlates with the Gross Motor Function Classification System (GMFCS) used for characterizing independent ambulation. Increasing EDACS level correlates with increasing risk for gastrostomy tube use (Benfer et al. 2017).

- Level I – Eats and drinks safely and efficiently.
- Level II – Eats and drinks safely but with some limitations to efficiency.
- Level III – Eats and drinks with some limitations to safety; there may be limitations to efficiency.
- Level IV – Eats and drinks with significant limitations to safety.
- Level V – Unable to eat or drink safely – tube feeding may be considered to provide nutrition.

Feeding interventions

The goal of feeding therapy is to aid families with providing nutrition in a manner that is manageable, safe, and efficient. Oral-motor therapy is an important part of feeding therapy to help the child develop and maximize eating skills. In order to optimize a child's oral potential in the home the following elements must be determined: safe and comfortable positioning, appropriate diet regarding consistency, and a supportive environment including appropriate applied techniques.

Oral-motor therapy

Oral-motor therapy, also known as sensorimotor treatment, is intended to augment oral-functional skills required for speech and eating. Of note, this is different from oral stimulation (see Applied techniques, on page 113). The skills addressed through oral-motor therapy include: awareness, strength, coordination, movement, and endurance of the lips, cheeks, tongue, and jaw as well as positioning and texture modifications. Oral-motor therapy may be beneficial in children with mild CP, but the scientific literature is equivocal regarding its benefits for children with severe CP (Gisel 1994, Gisel et al. 1995).

Positioning

The optimal position for children with feeding difficulties is one that provides good trunk and head stability with alignment at the hips and knees, and with the feet resting securely on a surface. This ideal positioning is nearly impossible when feeding a child in a lap, but can be achieved in most wheelchairs. It is usually easier to optimally position a child in the morning when he/she is rested; however, this may be more difficult after multiple transitions in and out of the chair throughout the day. Of note, care providers in the school system who are feeding children with oral-motor dysfunction should also have training in optimal positioning and feeding techniques.

Diet consistency

The most appropriate diet consistency for a child with CP will depend on the extent of the oral-motor dysfunction as well as the time of day, positioning, environment and fatigue level of the child. Due to the impact of limited lingual manipulation and chewing abilities, many children will require modified textures. Children with significant oral-motor dysfunction need a pureed diet that does not require any lateralization to the molars for chewing. Others may only require a soft or chopped diet. Most children with oral-motor dysfunction will have significant difficulty with drinking via cup or straw. Some children with severe oral-motor dysfunction may not be safe for any oral intake and require a gastrostomy tube for nutrition and hydration. Children with hypotonia may not transition to age-appropriate diet textures, but may be able to transition to increased textures over time.

It is useful to systematically characterize food and fluid consistency so that health care providers, therapists and caregivers can be consistent in describing a child's diet. The National Dysphagia Diet (NDD) classifies foods based on the following descriptions.

Solid textures:

- NDD level 1 (pureed): These foods require no chewing, are homogenous, very cohesive and are a pudding consistency. It is important to keep in mind that 'pureed' foods are not all equal. Thicker purees are usually recommended in order to prevent aspiration during the swallow as they move in a slow manner. Thicker purees, however, result in increased amount of food residue in the pharynx following the swallow, especially in children with hypotonia.
- NDD level 2 (mechanical altered): These foods require some chewing ability, but are cohesive, moist and semi-solid.
- NDD level 3 (advanced): These foods require some chewing ability, but are rarely multi-layered or mixed consistencies (e.g. vegetable soup).

Drinking is often difficult for children with oral-motor dysfunction because of the speed at which fluid flows. It can also be challenging for a child to pull liquids from a cup or straw. Maintaining adequate hydration can therefore be a slow and tedious process. Just as with solid food textures, a variety of liquid viscosities exist for children with oral-motor dysfunction.

Liquid viscosity:

- Thin liquids: Regular liquids such as water, juice or milk. Milk and chocolate milk as well as some prepared formulas tend to be slightly more viscous than water or juice.
- Nectar liquids: Thickened to the level of 'nectar' as found in canned fruit.

- Honey liquids: Thickened to the level of honey.
- Spoon-thick liquids: Resemble purees.

Environment

The ideal feeding environment is one that is quiet and calm, especially since some children with CP startle easily. Infants in general feed better before reaching extreme hunger, fatigue and related irritability. Infants with CP who become upset may demonstrate increased extensor tone, making feeding even more challenging. In older children the environment should be conducive to safe and efficient oral intake but should not be socially isolating if possible. Sometimes feeding children with complex CP in a crowded and noisy school cafeteria can be particularly challenging.

Applied Techniques

A few hands-on techniques may be utilized during feeding to improve efficiency and safety.

- **Jaw/cheek support** – The thumb and pointer finger of the person feeding are positioned on either side of mouth on the cheek surface and under the chin. This helps the child achieve mouth closure, which aids in triggering a swallow. This technique is most useful in children with hypotonia and/or mild hypertonia. In children with more extensive oral-motor dysfunction, it is not usually effective.
- **Lip closure** – This technique is applied in conjunction with jaw/cheek support to help the lips close. This maneuver assists in achieving a more efficient and effective swallow.
- **Direct food placement –** This technique varies from child to child depending on the level of oral-motor dysfunction. Some children are able to keep pureed foods in their mouths if it is placed on the sides of the tongue/cheek pocket rather than directly on the surface of the tongue. Other children can 'munch' dissolvable foods safely when they are placed directly on their molars.
- **Oral stimulation –** This is a theoretical strategy in which sensory stimulation prepares the child and their mouth for eating more efficiently and safely. For children with sensory processing issues, oral stimulation is another strategy, though the scientific evidence is limited and unclear as to whether it results in more efficient or safe eating for children with severe oral-motor dysfunction (Arvedson 2013).

Hunger and satiety

Hunger/satiety is a complex cycle regulated by numerous different elements including physiologic, developmental and environmental elements. Children with CP are at high

risk for disordered hunger/satiety regulation for many reasons. Impairments in motor activity with associated reduced metabolic needs results in decreased hunger. Differences in tone and gastro-intestinal motility cause constipation and gastroesophageal reflux disease (GERD), which negatively affect appetite. Lengthy mealtimes and sensory impairments also detract from the feeding experience. Supplemental feeding, such as with a feeding tube, often make it difficult for a child to have normal regulation of hunger/satiety cycles.

In addition to physiological differences, children with CP may be more easily influenced by the environments in which they eat. School cafeterias, birthday parties, sporting events may be too noisy and result in increased tone or startle responses. Caregiver stress due to concerns about inadequate nutrition and lengthy feeding times impacts feeding dynamics, especially in children with CP who already experience poor growth.

Regulating hunger/satiety patterns in children with CP should be a key component of the intervention plan. Children should be transitioned to age-appropriate feeding regimen if possible, especially when receiving gastrostomy tube feeds. Aggressive management of constipation, GERD and other gastro-intestinal dysmotility is important as well. Lastly, oral-motor skills should be optimized in order improve efficiency and safety.

Growth and nutrition

Many children with severe CP are inefficient eaters. These children may be able to chew and swallow safely, but may take excessive time to feed, and/or fatigue easily doing so. The disproportionate effort required to consume food ultimately limits nutrition and growth. Children who eat inadequate calories and protein at first may continue to gain weight, but at a suboptimal rate. When insufficient nutrition persists, linear growth and body mass composition are compromised as well.

Inadequate nutritional intake resulting in poor growth evolves over months to years (Stevenson and Conaway 2007). Infants with CP and associated oral-motor impairment may grow adequately in infancy as they rely on the primitive feeding reflex of sucking/swallowing. As infants develop, they require more calories but are unable to maintain appropriate growth during the transition to more complex oral-motor skills. Children who are inefficient eaters might also struggle with catch-up weight gain after recurrent illnesses. Other children with hyperkinetic CP might have a mismatch of calorie requirements and nutritional intake.

Although malnutrition can refer to both under- and over-nutrition, for the purposes of this handbook, malnutrition will refer only to under-nutrition. Prolonged malnutrition

leads to micronutrient deficiencies, low fat stores and decreased height velocity (Krick et al. 1996, Henderson et al. 2002, Stevenson and Conaway 2007). Inadequate nutrient intake negatively affects growth, development, and other relevant outcomes (Mehta et al. 2013). Other consequences of malnutrition include fatigue, increased illness/hospitalization, and familial stress regarding poor growth.

Oral-motor impairment with associated malnutrition is usually a contributing factor to poor growth among children with severe CP, though various other factors have been identified. (Henderson et al. 2005, Stevenson and Conaway 2007, Kuperminc et al. 2009). Other reasons for poor growth include neuroendocrine abnormalities, muscle-bone interactions, and psychosocial influences. Laboratory assessment may be required to identify medical causes of poor growth. Causes of poor growth to consider include hyper- or hypothyroidism, growth hormone deficiency, delayed puberty, and inflammatory disease.

Despite all known factors that might contribute to poor growth, children with CP overall grow differently compared to their typically developing counterparts. In general, children with CP are shorter and smaller compared to peers, and this trend is positively correlated with severity of GMFCS classification. Children with CP tend to have decreased fat mass, lean muscle mass and bone density (Stallings et al. 1995, Henderson et al. 2005). Poor growth, therefore, may be a marker of severe CP, and may not be modifiable in many cases.

When assessing the growth and nutrition of children with CP it is important to consider the complex interaction of multiple influences. Additionally, the differences in body mass composition, which relate to CP severity, should be taken into account. As such, the best strategy is a combination of tools to determine whether a child with CP is growing appropriately and is well nourished.

Growth assessment

Obtaining typical anthropometric measurements can be challenging due to commonly associated physical differences in children with severe CP. Children with CP often have significant scoliosis and contractures, which affect traditional length/height measurements. Weight can also be difficult to obtain using standard scales if the child is non-ambulatory. BMI is not an appropriate calculation using weight and height because children with CP have lower lean muscle mass. (Kuperminc and Stevenson 2008). This measurement may be helpful to assess trends and patterns, but should not be used alone as an indicator of nutrition status, if used at all (Stallings et al. 1995, Samson-Fang and Stevenson 2000). The criterion standard for assessing body composition (includes fat, water, protein and bone) is a Dual X-Ray Absorptiometry (DXA) scan. However, this is not readily available for clinicians so other anthropometric measures are available and acceptable (Kuperminc and Stevenson 2008).

Table 7.1 Equations for estimating height using segmental measurements in children with cerebral palsy under 12 years of age

Segmental Measure	Prediction equation for height (cm)	Standard error of the estimate (cm)
Upper arm length (UAL)	(4.25 x UAL) + 21.8	± 1.7
Tibial length (TL)	(3.26 x TL) + 30.8	± 1.4
Knee height (KH)	(2.69 x KH) + 24.2	± 1.1

Source: Samson-Fang and Bell 2013

Stature/height

Obtaining a height measurement can be challenging if not impossible due to physical differences (Kuperminc and Stevenson 2008). If the child is unable to stand, but is easily and reliably able to lie on a recumbent board, this is an acceptable alternative. If unable to obtain an accurate standing or recumbent length measurement, other options are available. Segmental measurements include ulnar length, knee height, and upper arm length (see Nutrition assessment, below for equations for estimating stature from these measurements). Knee height is the most reliable (Table 7.1). A flexible tape measure along the patient in a lying position is not a valid, reliable or reproducible method for obtaining a height measurement (Samson-Fang and Bell 2013).

Weight

As mentioned, the body composition of children with CP is different, and therefore weight does not reflect typical distribution of body fat and muscle (Stallings et al. 1995, Samson-Fang and Stevenson 2000, Kuperminc et al. 2010). Children with severe CP often appear low on the typical weight curve, but may have appropriate adiposity. Therefore, monitoring weight increase over time is useful. Consequently, it is extremely important to have accurate weight measurements in order to assess growth trends reliably. The minimal goal for children who are less than the 5th centile for age (CDC or WHO growth charts) is to follow his/her own curve and maintain consistent weight gain. Another useful resource are the California Growth Charts (Brooks et al. 2011).

Accurate weight measurements in infants require a naked weight. For children and adolescents who are non-ambulatory, the ideal is to weigh on a chair scale or wheelchair scale with only a light layer of clothing. If it is not possible to place the child on a scale independently, the child and caregiver can be weighed together and then subtract caregiver's weight (Samson-Fang and Bell 2013).

Triceps skinfold

This measurement provides useful information regarding fat stores and it is thought to be the best screening tool for malnutrition in this population (Samson-Fang and

Stevenson 2000). To obtain an accurate measurement, determine the midpoint between the top of the shoulder and the elbow. Then pinch the skin so that the fold is running vertically. Using a skinfold caliper, obtain the measurement (recommend obtaining 2–3 measurements to ensure accuracy). The CDC provides growth references for children from 1.5 to 20 years of age for plotting these measurements.

Triceps skinfold is a better indicator of fat stores compared to weight for length or BMI, as it measures fat stores directly. The goal is for these measurements to be greater than the 10th centile for age as it has been found that children with measurements less than the 5th centile are at higher risk for malnutrition (Samson-Fang and Stevenson 1998) and associated increased health care utilization and decreased social participation. It is important to note that some children with severe CP may hold their fat stores more centrally (in the abdominal cavity) and may demonstrate lower triceps skinfold measurements (Kuperminc et al. 2010, Gurka et al. 2010). Thus, triceps skinfold may underestimate total body fat stores.

Interpreting anthropometrics

Given the constraints of typical anthropometric measurements in children with CP, it is important to consider each measurement respective to other information and review growth trends. If a child is generally following his/her established curve for weight and length measurements, has adequate triceps skinfold (greater than 10th centile for age), and is not significantly limited in oral intake, it is appropriate to continue monitoring and re-assessing nutrition status every 6 months.

CP-specific growth charts are available (Brooks et al. 2011). It is important to note that these charts portray how children with CP *have* grown and not necessarily what is optimal growth (Kuperminc and Stevenson 2008, Krick et al. 1996). These growth charts are based on severity of gross motor impairment in children with CP, with higher level of impairment correlating with poorer growth. They are extremely useful as a clinical guide for children with complex CP. Another resource is for assessing growth is the online PediTools: Clinical Tools for Pediatric Providers which has growth calculators for weight, length/height, triceps skinfold, and mid-upper arm circumference (CDC and WHO), and provides centiles and Z-scores for age (Chou 2012; Available at www.peditools.org).

Nutrition assessment

When there are concerns about adequate nutrition, more frequent growth and diet assessments may be necessary to monitor trends or changes. Importantly, regardless of initial predictions of caloric needs, it is critical to assess weight gain and skinfolds over time. Consultation by a dietician may result in a calculated estimate of nutrition needs, using a variety of formulas and equations. Regardless, it is essential to monitor weight gain at any given estimate of calorie needs to determine if that level is excessive, inadequate or adequate. If a child is growing along his/her expected curve, this should

be considered appropriate growth(Kuperminc et al., 2013). Many equations require a weight, height, or both, and thus accurate measurements are requisite.

Assessing physical status is an important component of a nutritional evaluation. The appearance of hair, skin and nails often provides clues to nutritional state. Pallor or pale skin can indicate iron, folate, or cobalamin deficiency. Lesions or fissures in the oral cavity may show signs of vitamin B deficiencies. Edema can be associated with protein malnutrition. Pressure ulcers and poor wound healing may reflect inadequate calorie

Table 7.2a Harris Benedict equation (using basal energy expenditure + activity/stress level)[a, b]

For males:	B.E.E. = 66.5 + (13.75 x kg) + (5.003 x cm) – (6.775 x age)
For females:	B.E.E. = 655.1 + (9.563 x kg) + (1.850 x cm) – (4.676 x age)

Source: Stallings et al. 1996

[a]Stress factors range from 1.2 to 2 depending on activity level

[b]Comment on stress factor: Tone should be taken into account when factoring in activity level for a child with severe cerebral palsy as hypertonicity can increase energy burned even if activity level or movement is low. For a patient who is wheelchair bound without much movement and without significantly increased tone, a provider may choose to use 1–1.2 as an activity factor, but should monitor weight gain closely. Clinical judgment plays a large role when determining activity factor in patients with severe cerebral palsy.

Table 7.2b Equation using kilocalories per centimeter in estimated or actual height

Children 5–11 years	Ambulatory = 14 kcal/cm
	Non-ambulatory = 11 kcal/cm

Source: Adapted from Weston 2012

Table 7.2c The World Health Organization equations for estimating resting energy expenditures

Age (years)	Resting energy expenditures (REE) (kcal/day)
0–3	Males: (60.9 x weight [kg]) – 54 Females: (61.0 x weight [kg]) – 51
3–10	Males: (22.7 x weight [kg]) + 495 Females: (22.5 x weight [kg]) + 499
10–18	Males: (17.5 x weight [kg]) + 651 Females: (12.2 x weight [kg]) + 746

Source: WHO 1985

and protein intake, or vitamin C, vitamin D and zinc deficiencies. Vitamin A deficiency may take the form of corneal clouding or ocular xerosis, whereas thiamin deficiency may present as nystagmus or ophthalmoplegia. Inadequate fat mass secondary to inadequate calorie intake may result in pubertal delays (2013, Pogatschnik 2011) (Table 7.3).

Laboratory assessment

Blood work may also be indicated as part of a complete assessment for growth and nutrition concerns. First consider other factors that might contribute to poor growth such as celiac disease, endocrine disorders, cystic fibrosis, etc. (Samson-Fang and Bell 2013).

Table 7.3 Physical findings and symptoms associated with malnutrition

	Physical exam and symptoms	Possible nutritional deficiency
Skeletal system	Epiphyseal enlargement of wrists, legs, knees; bowed legs; frontal bossing of the head; bone pain	Vitamin D
Muscular system	Wasted appearance	Protein calorie
	Weakness	Phosphorous
		Potassium
		Vitamin D
		Vitamin C
		Vitamin B6 (pyridoxine)
	Absent deep tendon reflexes, foot and wrist drop	Vitamin B1 (thiamin)
	Muscle cramps	Vitamin D
		Calcium
		Magnesium
		Potassium
		Chloride
	Muscle pain	Vitamin D
		Vitamin B7 (biotin)
Nervous system	Inability to concentrate, memory impairment, disorientation, irritability	Vitamin B1 (thiamin) Vitamin B9 (folic acid) Vitamin B12 (cobalamin)

Table 7.3 (Continued)

	Physical exam and symptoms	Possible nutritional deficiency
	Seizures, behavioral disturbances	Vitamin D
		Calcium
		Magnesium
		Zinc
	Peripheral neuropathy (numbness/ tingling and weakness)	Vitamin B1 (thiamin)
		Vitamin B5 (pantothenic acid)
		Vitamin B6 (pyridoxine)
		Vitamin B9 (folic acid)
		Vitamin B12 (cobalamin)
		Phosphorous
		Copper
Eyes	Angular blepharitis (inflammation of the eyelids)	Vitamin B2 (riboflavin)
		Vitamin B6 (pyridoxine)
		Vitamin B7 (biotin)
		Zinc
	Pale conjuctiva	Vitamin B6 (pyridoxine)
		Vitamin B9 (folic acid)
		Vitamin B12 (cobalamin)
		Iron
		Copper
	Keratomalacia (opaque/dull cornea, night blindness), Bitot spots (white or grey spots on conjunctiva)	Vitamin A
	Ophthalmoplegia	Vitamin B1 (thiamin)
		Phosphorous (associated with rickets)

Table 7.3 (Continued)

	Physical exam and symptoms	Possible nutritional deficiency
Integumentary system		
A) Skin	Pallor	Vitamin B9 (folic acid)
		Vitamin B12 (cobalamin)
		Iron
		Vitamin C
	Dark cheeks and under eyes, scaling skin around nostrils	Protein calorie
		Vitamin B2 (riboflavin)
		Vitamin B3 (niacin)
		Vitamin B6 (pyridoxine)
	Pellagra (thick, dry, scaly hyperpigmentation on sun-exposed areas)	Vitamin B3 (niacin)
		Vitamin B6 (pyridoxine)
		Tryptophan
	Yellow pigmentation	
		Vitamin B12 (cobalamin)
	Follicular hyperkeratosis	Vitamin A
		Vitamin C
	Seborrheic dermatitis (scaly, waxy, crusty plaques on scalp, nasolabial folds, lips)	Vitamin B2 (riboflavin)
		Vitamin B6 (pyridoxine)
		Vitamin B7 (biotin)
		Vitamin A
		Zinc
		Essential fatty acid deficiency
	Eczema	Vitamin B2 (riboflavin) Zinc
	Xerosis (abnormal dryness)	Protein calorie Vitamin A

Table 7.3 (Continued)

	Physical exam and symptoms	Possible nutritional deficiency
	Slow wound healing, decubitus ulcers	Protein calorie Vitamin C Zinc
	Petechia (purple/red dots) or purpura (purple spots/patches) on skin	Vitamin C Vitamin K
	Cellophane appearance of skin	Protein calorie Vitamin C
	Edema	Protein calorie
	Poor skin turgor	Dehydration
B) Hair	Alopecia (thin, sparse hair)	Protein calorie Zinc Iron
	Easily plucked without pain, dull/dry/lack of shine	Protein calorie Essential fatty acid
	Depigmentation, color changes	Protein calorie Manganese Selenium Copper
	Flag sign (alternating color on hair strand due to lack of melanin)	Protein calorie
	Lanugo (find, soft hair)	Malnutrition
	Cork-screw hair and/or peri-follicular hemorrhage	Vitamin C
C) Nails	Brittle, soft, thin	Severe calorie Vitamin A Magnesium Selenium
	Central ridges	Protein calorie Vitamin B9 (folic acid) Iron
	Beau's lines (transverse ridges or horizontal grooves)	Protein calorie Zinc Calcium

Table 7.3 (Continued)

	Physical exam and symptoms	Possible nutritional deficiency
	Koilonychia (spoon-shaped or concave)	Protein Vitamin B9 (folic acid) Vitamin B12 (cobalamin) Iron
	Muehrcke lines (transvers white lines)	Malnutrition
Mouth and oral cavity	Soreness, burning mouth, tongue, lips	Vitamin B2 (riboflavin)
	Angular stomatitis or chelitis (redness or fissures at corners of mouth)	Vitamin B2 (riboflavin) Vitamin B3 (niacin) Vitamin B6 (pyridoxine) Vitamin B12 (cobalamin)
	Glossitis (sore, swollen, beefy-red tongue)	Vitamin B2 (riboflavin) Vitamin B3 (niacin) Vitamin B6 (pyridoxine) Vitamin B9 (folic acid) Vitamin B12 (cobalamin)
	Gingivitis (swollen, spongy gums that bleed easily)	Vitamin B3 (niacin) Vitamin B9 (folic acid) Vitamin C Vitamin D Zinc
	Missing teeth or late eruption	Malnutrition
	Dental caries	Fluoride Vitamin D Vitamin B6 (pyridoxine)
Gastro-intestinal system	Anorexia, flatulence, diarrhea	Vitamin B6 (pyridoxine) Vitamin B12 (cobalamin)

Children with severe CP are at-risk for poor bone mineral density and osteoporosis, therefore regular assessment of vitamin D (25-OH), alkaline phosphatase, calcium and phosphorus is recommended (Fehlings et al. 2012). C-reactive protein is a useful indicator of inflammation which may lead to increased calorie requirements (Samson-Fang and Bell 2013). A complete blood count helps with determining anemia. If significant concern for malnutrition and/or dehydration is present, consider a basic metabolic panel to assess for electrolyte disarray and/or renal

Table 7.4 Basic laboratory assessment for malnutrition

Anemia	Complete blood count
Bone health	Vitamin D 25-OH Calcium Alkaline phosphatase Phosphorous
Hydration status and re-feeding syndrome risk	Complete metabolic panel Magnesium Phosphorous

failure. Decreased albumin may reflect chronic malnutrition; however, albumin levels are affected by inflammation and fluid shifts, thus is not usually useful (Samson-Fang and Bell 2013). If available to a clinician, DXA allows for assessment of body composition including fat and lean mass stores (Samson-Fang and Bell 2013) (Table 7.4).

Interventions for poor growth and malnutrition

Children with severe CP who have malnutrition are often amenable to several fairly simple interventions. It is ideal to involve a dietician or gastroenterologist, but this is not uniformly possible. It is also useful to have a speech/language or occupational therapist assess feeding to ensure the child is consuming a diet of appropriate consistency to be safe and most efficient.

An essential component for increasing daily calorie intake is implementation of a meal schedule in order to provide the child with multiple eating opportunities (3 meals plus 2–3 snacks). The caregiver should incorporate nutrient rich foods at each eating occasion. Sources of nutrient rich foods include proteins (meats, fish, beans, eggs), fruits, vegetables, whole grains and dairy.

Oral nutrition supplements or 'calorie boosters' to increase calorie/protein/micronutrient intake are often necessary. Calorie boosters, such as heavy cream, whole milk, dry milk powder, butter, cheese, peanut butter, avocado and black beans, are easy foods to add to the diet. Caregivers can create 'milkshake' supplements using ice cream, milk, fruits, and vegetables to increase calories in liquid consistency. The other option is to use commercially available oral supplementation. See Table 7.5 for a list of common pediatric and adolescent oral supplementation (not an all-inclusive list). A variety of oral nutritional supplements exist and determining which supplement is most appropriate often requires the expertise of a nutritionist. A growing trend has emerged in the U.S. and elsewhere regarding the use of blended table foods in place of commercial

Table 7.5 Common pediatric and adolescent oral supplementation

Pediatric specific supplements *Generally are interchangeable*	Adult/adolescent supplements
Meal Replacements: Pediasure (children 2–13 years of age) • Multiple flavors • Milk based, but suitable for lactose intolerance Pediasmart • Powdered version of Pediasure • Not as easily found in stores Carnation Instant Breakfast • Multiple flavors • Powder that is to be mixed with 8oz milk • Usually a cheaper option to Pediasure Bright Beginnings Soy Parent's Choice Pediatric Drink (Walmart brand) Store brands that are comparable to the above	**Meal Replacements** Ensure or Boost Products • Variety of calorie levels (150–350 calories per container) depending on the level of support needed from supplement Carnation Instant Breakfast • Multiple flavors • Powder to be mixed with 8oz milk • Usually a cheaper alternative to Ensure/ Boost Store brands comparable to the above **Non-nutritionally complete supplements** Ensure Clear or Boost Breeze • Juice-based supplements that provide additional calories and protein, but do not contain fat • Not to be used as a meal replacement, but can be used if diet is below adequate, however patient is able to take most nutrients orally and needs small amounts of supplementation • Can be used in the pediatric population as needed

formulas. To insure the adequacy of macro and micronutrients, we recommend that the use of homemade blended "formulas" be supervised by a nutritionist.

Micronutrient supplementation may be indicated if laboratory assessment indicates deficiency or if intake is obviously below recommended daily intake. Vitamin D and calcium supplementation are frequently necessary in children with CP due to several risk factors for poor bone health. If general nutrition is a concern, providers should recommend a pediatric multivitamin supplement.

The decision to place a gastrostomy tube in a child with CP should be made after careful consideration of safety and efficiency of oral intake, growth patterns, number/ severity of respiratory illnesses, and overall quality of life. Many children with CP have oral-motor skills that detract from pleasant mealtime interactions due to concerns for safety and quantity of nutrition. This can result in stress for the family and direct health

concerns for the children. Healthcare professionals should work with the family and other service providers to achieve the most pleasant, safe and efficient manner of eating for children with CP. Placement of a gastrostomy tube does not preclude oral feeding but may optimize growth, allow for accurate delivery of medication and fluids, and may offer additional time for other aspects of life.

Obesity

Obesity can occur in children with severe CP, especially in those who are gastrostomy tube-dependent. Children who are fed via gastrostomy do not have the ability to self-regulate intake and often have low metabolic needs. These children might easily receive calories in excess and gain weight rapidly, especially in the immediate period after placement of a feeding tube. Other children may have low resting metabolic needs due to decreased muscle mass in addition to low energy needs from lack of movement. At times it is difficult to provide these children with adequate micronutrients due to substantially low caloric needs.

Consequences of obesity in this population include metabolic syndrome and related issues. Obstructive sleep apnea is another serious consideration since these children are already at higher risk due to their motor impairment. Furthermore, it is more difficult for caregivers to move and care for overweight children with severe CP. To monitor children with severe CP at-risk for obesity, track weight gain velocity and triceps skinfold measurement trends over time, at regular intervals. Concerns should arise with consistent crossing of centiles in weight and triceps skinfolds (specifically when TSF reaches greater than the 85th centile for age).

Recommendations to deter obesity include decreased portion sizes, high-calorie foods, and carbohydrate-laden beverages. If the child receives tube feeds, slowly reduce calories, and closely monitor weight and triceps skinfold changes. Micronutrient supplementation may be required if calorie needs are extremely low. The goal is to maintain adequate linear growth and prevent further weight gain, or promote slow weight loss if no more linear growth is to occur.

Conclusion

Children with severe CP often require an extraordinary amount of basic care, with feeding difficulties and the related nutrition challenges comprising a large portion of their medical needs. It is critical to address feeding difficulties in these children not only to optimize overall health and growth, but also to potentially decrease family stress related to feeding. Although children with severe CP may struggle with oral-motor impairment

and related nutrition/growth problems, various strategies exist to help optimize the safety and efficiency of feeding. Ideally, this may potentially result in increased pleasurable mealtime experiences for the child but also improved family harmony overall. Children and their families benefit from positive mealtime socialization and it is the job of the health care team to facilitate this experience by providing appropriate guidance, monitoring and interventions.

Key points

- Most children with complex CP struggle with feeding difficulties, although the degree of oral-motor impairment may vary widely with some children requiring only prolonged feeding times and others a gastrostomy tube.
- It is important to assess for safety and efficiency of eating, as well as the impact of the feeding difficulties on the family dynamic.
- Various strategies for improving safety and efficiency of feeding exist and include oral-motor therapy, diet consistency, positioning and environmental accommodations, and applied techniques.
- Children with severe CP overall grow differently compared to their typically developing counterparts. In general, children with CP are shorter and have decreased fat mass, lean muscle mass and bone density. As such, traditional strategies for assessing growth and nutrition require adaptation.
- Obesity among children with complex CP is commonplace due to multiple factors. Children with a gastrostomy must be monitored closely to prevent unintended obesity.
- For many children with severe CP, consultation with nutrition and a speech-language therapist is recommended and helpful.

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NOTE: Figures, boxes, tables and appendices are denoted by an italic, lower case *f, b, t* and *app* respectively. "Cerebral Palsy" is abbreviated to "CP" in subheadings throughout.