Pediatr Gastroenterol Hepatol Nutr. 2019 Mar;22(2):122-131
https://doi.org/10.5223/pghn.2019.22.2.122
pISSN 2234-8646·eISSN 2234-8840

Pediatric Gastroenteology, Hepatology & Nutrition
pghn

Review Article

Evaluation and Treatment of Malnutrition and Associated Gastrointestinal Complications in Children with Cerebral Palsy

Ivana Trivić[1] and Iva Hojsak[1,2,3]

[1]Referral Center for Pediatric Gastroenterology and Nutrition, Children's Hospital Zagreb, Zagreb, Croatia
[2]University of Zagreb, School of Medicine, Zagreb, Croatia
[3]Josip Juraj Strossmayer University of Osijek, School of Medicine, Osijek, Croatia

Received: Dec 6, 2018
Accepted: Dec 31, 2018

Correspondence to
Iva Hojsak
Referral Center for Pediatric Gastroenterology and Nutrition, Children's Hospital Zagreb, Klaićeva 16, Zagreb, Croatia.
E-mail: ivahojsak@gmail.com

Conflict of Interest
Ivana Trivić has no financial conflicts of interest. Iva Hojsak received honorarium for lectures or consultation for BioGaia, Nutricia, Nestle, GM pharma, Chr Hansen.

ABSTRACT

The majority of children with cerebral palsy (CP) have feeding difficulties and are especially prone to malnutrition. The early involvement of a multidisciplinary team should aim to prevent malnutrition and provide adequate nutritional support. Thorough nutritional assessment, including body composition, should be a prerequisite for the nutritional intervention. As in typically-developed children nutritional support should start with dietary advice and the modification of oral feeding, if safe and acceptable. However, for prolonged feeding, in the presence of unsafe swallowing and inadequate oral intake, enteral nutrition should be promptly initiated and early gastrostomy placement should be evaluated and discussed with parents/caregivers. Gastrointestinal problems (oropharyngeal dysfunction, gastroesophageal disease, and constipation) in children with CP are frequent and should be actively detected and adequately treated as they can further worsen the feeding process and nutritional status.

Keywords: Children; Cerebral palsy; Malnutrition; Nutritional assessment; Diet therapy

INTRODUCTION

Cerebral palsy (CP) comprises of a heterogeneous group of early-onset, non-progressive, neuromotor disorders which affect the developing fetal or infant brain [1]. The latest systematic review and meta-analysis, published in early 2013, estimated the prevalence of CP to be 2.11 per 1,000 live births and as high as 59.18 per 1,000 live births among neonates weighing less than 1,500 g. The prevalence has remained constant over the recent decade, despite the increased survival of at-risk preterm infants [2]. The life expectancy of children with CP has gradually improved and thus, the prevalence and consequences of feeding difficulties are on the rise [3]. Based on data from the North American Growth in CP Project, 58% of children with moderate to severe CP had feeding difficulties, in which 23% were severe [4]. Feeding disorders play an important role in the development of malnutrition, documented in 29%–46% of CP children. The prevalence of undernutrition increased with older age, lower intelligence quotients, and more severe neurological impairment [5]. Beside growth failure, the most evident consequence of malnutrition, other include decreased cerebral function and reduced potential for development, impaired immune function, impaired circulation with poor wound healing, diminished respiratory muscle strength [3].

The importance of malnutrition and the need for nutritional management have been recognized by many societies. Most recently, the European Society for Pediatric Gastroenterology Hepatology and Nutrition (ESPGHAN) published guidelines for the evaluation and treatment of gastrointestinal and nutritional complications in children with neurological impairment [6].

The aim of this article was to review the literature on the assessment of nutritional status and nutritional management in children with CP.

NUTRITIONAL ASSESSMENT

Nutritional assessment in children with CP would ideally be performed by a multidisciplinary team consisting of a physician (preferably pediatric gastroenterologist), dietitian, nurse, speech and language therapist, physical therapist, occupational therapist, and psychologist [6]. Early integration of a multidisciplinary team should aim to identify children at risk for malnutrition and to provide nutritional management before malnutrition develops. As with any nutritional assessment, it should begin with a detailed clinical history and physical examination, followed by anthropometrics (methods assessing body composition), and laboratory methods [6].

Physicians should obtain information regarding the etiology and severity of the child's neurological impairment, as well as comorbidities and prescribed medications [7]. Information relating the child's motor ability should be objectified using the Gross Motor Function Classification System (GMFCS) and combined with information relating to psychomotor development, communication skills, as well as functional vision. The inability to communicate, together with moderate to severe cognitive impairment, may affect a child's ability to convey thirst, hunger and satiety, as well as to express preferences regarding food texture, flavor, and any discomfort during feeding. Other factors which can negatively influence are the visual impairment, scoliosis and contractures causing poor positioning [7]. Furthermore, antiepileptic drugs can reduce alertness and cause a variety of gastrointestinal adverse effects, such as heartburn, nausea, vomiting, gum problems, diarrhea, and constipation [8]. Suboptimal dosing with antiepileptic drugs and frequent seizures can negatively influence the feeding, as well [7].

It is very important to properly assess nutritional intake. To increase the accuracy, 24-hour food recall and/or a 3-day food diary can be used to provide the necessary information. Dietitians are advised to be cautious when interpreting diaries since families may overestimate the amounts consumed by overlooking spillage or vomiting and underestimate the time required for feeding [7]. Since feeding and nutritional problems are more likely to occur in children with severe motor disabilities [9], full assessment in such cases necessitates the evaluation of oral motor skills and swallowing by other professionals, such as speech and occupational therapists [7]. The best way to gain insight into the feeding process is either to have meal time at home video recorded by parents or to observe feeding in a hospital/outpatient setting.

In the physical examination, special attention should be given to the inspection of the skin and peripheral circulation. Poor wound healing, the presence of decubitus ulcers, cold extremities, prolonged capillary refill time, and signs of micronutrient deficiencies can indicate malnutrition [6].

https://doi.org/10.5223/pghn.2019.22.2.122

Anthropometrics

Anthropometric assessment is a widely used method for assessing growth and nutritional status. In children with CP, measurement can be difficult and the references commonly used in pediatrics tend to misinterpret malnutrition in this specific group of patients [10].

Even weight measurements often require special scales, like wheelchair scales, sitting scales, and hoist scales [11]. Height measurement is even more challenging. Standing height or supine length can be used in children who can stand or lay down straight. However, in children who are unable to stand upright due to scoliosis, limb contractures, and spasticity, alternative measurements for the height assessment should be segmental lengths, such as knee-heel length, tibia length, and ulnar length, assessed by sliding calipers [6]. Special equations or charts can then be used to calculate the standing height. The anthropometric measurements have proper value if plotted into adequate growth charts. Several CP-specific growth charts containing estimated weight-for-age or height-for-age percentiles have been created. Most recently, in 2011, Brooks et al. [12] published clinical growth charts for children with CP, stratified according to gender and GMFCS level. However, these charts show how children with CP are growing, not necessarily how they should grow. Therefore, ESPGHN recommends the use of growth charts for typically-developed children to assess growth in children with CP [6].

Regardless, nutritional status in children with CP should not be estimated solely by weight and height measurements. It should also depend on follow-up and body composition assessments [6].

Body composition assessment

According to the ESPGHAN recommendation, children with neurological impairments should have their body composition assessed [6] and different methods have been used to assess it. The measurement often referred to as the criterion standard for the assessment of body composition is whole-body dual-energy x-ray absorptiometry (DXA) [13]. DXA can detect known alterations in body composition in children with CP, such as increased total body water, decreased fat-free mass, and decreased bone mineral density [14]. However, DXA scanning can often be challenging in children with CP due to severe scoliosis, joints contractures, and poor positioning. Other methods used to determine body composition include deuterium oxide (D2O) dilution, underwater weighing, and bioelectrical impedance (BIA). Several studies have evaluated the role of BIA in the body composition assessment and found that BIA estimated body composition well compared to standard methods, such as DXA and D2O dilution [15,16]. Thus far, even more cost-effective and widely-available methods are the measurement of skinfold thicknesses, primarily the triceps and subscapular skinfold thicknesses. The results can then be evaluated by different equations, mainly the Slaughter equation for typically-developed children [17]. Though, these equations do not take into account the different body compositions of children with CP, in whom more body fat is stored centripetally and, consequently, so their total body fat percentage can be underestimated [13]. In order to improve the accuracy of the Slaughter equation, Gurka et al. [18] developed coefficients that correct for sex, race, size, pubertal status, and GMFCS level, minimizing the errors.

Laboratory methods

There is no single laboratory marker representing adequate or inadequate nutritional status. Studies evaluating the micronutrient status of children with CP found that deficiencies for

https://doi.org/10.5223/pghn.2019.22.2.122

iron, zinc, copper, vitamin D, carnitine, folic acid, and vitamin B12 were common and their incidence ranged between 10% and 55% [19,20]. The assessment of micronutrient status, as part of the assessment of nutritional status in children with CP, therefore, is strongly encouraged [6].

Definition of undernutrition

As already mentioned, the determination of nutritional status in CP children can be challenging. Therefore, ESPGHAN recommends the use of 1 or more red flag warning signs, including physical signs of undernutrition (like pressure sores and poor peripheral circulation), weight for age *z* score <−2, triceps skinfold thickness in the <10th centile for age and sex, mid-upper arm circumference or muscle area in the <10th percentile, faltering weight, and/or failure to thrive [6].

NUTRITIONAL REQUIREMENTS

There are no appropriate specific recommendations for assessing the energy requirements in children with CP [5]. Neurological impairment in children with CP varies greatly, and therefore, nutritional requirements in those children cannot be generalized. Children with CP who can walk require more energy for walking, while those dependent on a wheelchair require 60% to 70% of the energy required by typically-developed children [21,22]. Therefore, nutritional requirements should be assessed individually. The best predictor of energy needs has been found to be fat-free mass [23]. Ideally, indirect calorimetry can be used to assess such needs [5], however, in regular clinical practice, this is difficult, time-consuming, and in many places, not available. Thus, most clinicians use the Schofield equation and dietary reference intake for typically-developed children [6]. These references should only be a starting point and regular follow-up is needed in order to tailor future nutritional intervention [6].

DIETETIC MANAGEMENT

The most appropriate mode of nutritional intervention should be determined based on the patient's age, clinical condition, gastrointestinal function, safety and feasibility of oral intake, dietary habits, and costs [24]. Although oral feeding is preferred, it should be advised if nutritionally sufficient, safe, stress-free, and if feeding time does not exceed 3 to 4 hours per day [6]. Optimization of oral intake can be done by increasing the energy content through the addition of energy-rich foods and textural changes [25]. In cases where nutritional requirements cannot be met orally, the use of enteral nutrition should be considered [24]. It needs to be emphasized that enteral tube feeding should before undernutrition develops if the child is at high-risk for malnutrition [6]. The choice of enteral formula should be based on the child's age, energy requirements, and mode of enteral access (**Table 1**). Blenderized or pureed food are often preferred by caregivers as they perceive benefits from giving food in its natural state and providing food eaten by other family members [26]. There is limited evidence showing that the prevalence of vomiting and use of acid-suppressive agents were significantly decreased or gagging/retching was reduced following Nissen fundoplication after blenderized tube feeding was initiated [27,28]. However, there is a concern regarding the nutritional adequacy and safety of home-prepared tube food [29]. Therefore, the ESPGHAN position paper advises caution and surveillance when blenderized diets are used for enteral feeding [6].

https://doi.org/10.5223/pghn.2019.22.2.122

Table 1. Type of enteral nutrition based on ESPGHAN recommendations [6]

Population	Type of feeds
Infants	Breastmilk, regular formula or nutrient dense infant formula if indicated
Children above 1 year	Standard polymeric (1 kcal/mL) age appropriate formula with fiber
Children with an increased energy requirements or poor volume tolerance	High energy formula (1.5 kcal/mL) age appropriate formula with fiber or dietary supplementation with glucose polymers and/or long-chain triglycerides to increase calorie intake
Severely undernourished children	The addition of protein (2.0–2.4 g/kg/day) and energy (additional 20% increase in energy intake)
Maintenance of enteral tube feeding in immobile children	Low fat, low calorie, high fiber, and micronutrient sufficient

ESPGHAN: European Society for Pediatric Gastroenterology Hepatology and Nutrition.

In children with CP, enteral nutrition is usually required for prolonged times, therefore, many centers offer early gastrostomy placement. Although early gastrostomy placement is frequently refused by parents in the beginning, a systematic review showed that this strategy significantly improved weight and height gain, overall nutritional status, subcutaneous fat stores, ease of feeding and, importantly, caregiver satisfaction [30]. Most frequently, gastrostomy tubes are placed endoscopically as percutaneous endoscopic gastrostomies (PEGs). However, children with CP often have significant scoliosis and PEG cannot be safely placed because transillumination of the stomach is often not visible. In these cases, a laparoscopic gastrostomy or laparoscopically-assisted PEG can be safe alternatives. Laparoscopic gastrostomies or laparoscopically-assisted PEGs have been shown to significantly lower the rate of major complications in these patients [31].

In some patients, gastrostomy feeding is not tolerated and post-pyloric or jejunal feeding can be recommended as an alternative. These cases include the presence of severe gastric motility disorder, severe gastroesophageal reflux disease (GERD) which cannot be managed by anti-reflux surgery, or in patients at very high-risk for aspiration [32]. A jejunal tube (nasojejunal tube or jejunal tube introduced through a gastrostomy or surgical transcutaneous jejunostomy) should be positioned distal to the Treitz ligament to prevent retrograde filling of the stomach [6,24].

Enteral tube feeding in the stomach should be administered as a bolus feeding if tolerated, however, children with high-caloric needs or with poor volume tolerance could benefit from a combination of overnight continuous feeding with boluses during the day [5,6]. Jejunal feeding should be provided as continuous iso-osmolar feeding [6].

GASTROINTESTINAL PROBLEMS IN CHILDREN WITH CP

The most prevalent gastrointestinal difficulty in children with CP is oropharyngeal dysfunction (OPD). OPD, including oropharyngeal dysphagia and oral motor dysfunction, is characterized by the presence of disturbances in 1 or more of the 3 phases of swallowing (oral, pharyngeal, and esophageal) [6]. The prevalence of OPD in children with CP is high, up to 90% [33], and it is more frequent in younger children and children with poorer gross motor function (GMFCS III to V) [34]. The possible clinical signs of OPD include sialorrhea, coughing, wet breathing, gagging, choking, and alteration in appetite. The assessment of OPD includes direct observation of meal times by appropriately trained professionals (speech therapist, physicians, nurses, dietitians, or others) [6]. Several scoring systems have been proposed for the assessment of OPD, but the Schedule Oral Motor Assessment (SOMA) and the Dysphagia Disorders Survey (DDS) are the tools are most widely used to support clinical decision-making [35]. The SOMA is mainly a test of oral phase dysfunction and is

https://doi.org/10.5223/pghn.2019.22.2.122

intended for use in children with CP aged between 10 and 42 months, while the DDS evaluates oral, pharyngeal, and esophageal phases and is designed for children with developmental disabilities between 3 and 13 years of age [35]. Finally, videofluoroscopy (VFS) is the key tool for the assessment of OPD [6]. It can be used to identify reduced lip closure, inadequate bolus formation, residue in the oral cavity, delayed triggering of pharyngeal swallow, reduced larynx elevation, coating on the pharyngeal wall, delayed pharyngeal transit time, multiple swallows, discoordinate pharyngeal motility, and silent aspiration [36,37]. In cases of high suspicious of an abnormal pharyngeal phase of swallowing, and when VFS founding are normal, high-resolution esophageal manometry can be recommended [6]. Management of OPD in children with CP requires a multidisciplinary approach. Although evidence regarding the treatment of OPD is limited, management should aim to optimize oral intake in cases where intake has been proven safe. Interventions, according to ESPGHAN, include speech and language interventions and/or the consistency of feeding modifications [6]. The highest rate of improvement was seen in children in whom only the oral phase of swallowing was affected, while a recently published study found low GMFCS to be an additional positive factor [34].

Up to 70% of children with CP suffer from GERD [38]. Due to the very high frequency of GERD and fragility of children with CP in whom diagnostic tests are difficult to perform, a trial of proton pump inhibitors (PPIs) with careful clinical follow-up is acceptable according to ESPGHAN recommendations [6].

However, persistent symptoms do require additional diagnostic tests, including esophageal pH-metry, multichannel intraluminal impedance, and/or upper endoscopy, to evaluate GERD. In some children with persistent vomiting, additional diagnostic workup (including barium swallow and abdominal ultrasound) can be recommended in order to exclude other problems causing intestinal obstruction [6].

The main treatment for GERD in children with CP is PPIs but that treatment can be combined with lifestyle changes. Lifestyle changes consist of adequate positioning and certain dietary modifications, including the thickening of liquid formula and selection of whey-based, instead of casein-based, enteral formula [39]. Therapy with PPIs effectively reduces acid reflux and, therefore, treats esophagitis, however it does not have an effect on the volume and the number of the reflux episodes [40]. Therefore, although not routinely prescribed, the use of prokinetic agents, primarily baclofen, can be recommended for children in whom other pharmacological treatments have failed [6,41]. Frequently, fundoplication needs to be considered in children with GERD which cannot be pharmacologically controlled [6,41]. There is always a question as to whether fundoplication should routinely be performed in children with CP who require gastrostomy placement. This question was mainly raised by some studies that found that PEG placement increased GERD episodes [42], however, the majority of data from other studies did not confirm this association [43,44]. Therefore, the combined procedure should be recommended only in selected patients.

Another very frequent gastrointestinal symptom in children with CP is constipation, at a prevalence of 26% to 74% [9,45]. The causes of constipation in disabled children include neuromuscular factors, such as intestinal motility disorders, hypotonia, skeletal muscle discoordination, and skeletal deformities, combined with prolonged immobility of the child, as well as nutritional factors, such as low fiber and poor fluid intake and pharmacological factors (e.g., anticholinergics and opiates have negative effects on intestinal and colonic transit time) [46]. Diagnosis and treatment should be the same as for typically-developed

https://doi.org/10.5223/pghn.2019.22.2.122

children [6]. Although more than 50% of children with CP chronically use laxatives, they are less responsive to treatment than typically-developed children so the dose needs to be adjusted [47]. Since fiber and fluid intake is inadequate in 53% of children with neurological impairments [48], the increase of daily fiber and fluid intake can be an additional strategy [6]. In cases of refractory constipation, anterograde continence enemas have been reported to be effective treatment options [49], but should be recommended only in cautiously-selected patients.

CONCLUSION

The prevalence of feeding difficulties and undernutrition is still high in children with CP. Early involvement of a multidisciplinary team should aim to detect children at risk for malnutrition and provide adequate nutritional support in order to prevent undernutrition. Thorough nutritional assessment, including body composition, should be a prerequisite for the nutritional intervention. Individual approaches, as well as early enteral nutrition, if required, should be applied and gastrointestinal problems should be recognized in a timely fashion and treated.

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