Review Article

Dysphagia in cerebral palsy

Annamaria Salghetti[*], Andrea Martinuzzi

E. Medea Scientific Institute, Conegliano Research Centre, Italy

Abstract. Feeding problems are often present in children with neuromotor impairment: dysphagia is usually seen in the most severe form of cerebral palsy and it's defined as the difficulty with any of the four phases of swallowing. Clinical consequences are malnutrition and recurrent chest infections that reduce expected duration and quality of life. In order to prevent these consequences it's important to detect with clinical and instrumental examinations dysphagia symptoms and to treat them. Clinical evaluation focuses mainly on the oral stage of dysphagia i.e. patterns of oral dysfunction but is not able to assess accurately the pharyngeal and esophageal phases that can be studied with instrumental evaluation like videofluoroscopy. Videofluoroscopy data provide the basis for an objective planning of the treatment only if combined with careful clinical examination. Treatment options include rehabilitative measures such as postural management and food texture modification and in the most severe cases surgical procedures.

Key words: Cerebral palsy, dysphagia

1. Introduction

Cerebral palsy (CP) is a well-recognized neurodevelopment condition beginning in early childhood and persisting through a person's lifespan. The most recent definition (1) describes CP as a group of disorders concerning the development of movement and posture, causing activity limitation, that are attributable to non-progressive disturbances that occur in the developing infant or fetal brain (1). The incidence of CP is estimated in 2-3/1000 live births (2). The motor disorders of CP are often accompanied by disturbances of sensation, cognition, communication, perception, behavior and epilepsy. The severity of disturbances varies greatly among subjects. Difficulties in the complex coordinated activity of feeding may be present above all in the most severe forms of CP (quadriplegia). Dysphagia is a swallowing problem defined as the difficulty with any of the four phases of swallowing: oral preparation, oral stage, pharyngeal stage or esophageal stage.

The prevalence of feeding problems in children with neuromotor impairments is very high. A community-based survey demonstrated oral motor dysfunction in more than 90% of a cohort of 49 children with CP (3). In spastic CP, the central nervous system (CNS) injury involves the corticobulbar and/or corticospinal tracts. Children with damage to the corticospinal and/or corticobulbar tracts can be expected to have greater dysfunction of those phases of swallowing that have significant voluntary components. The phases of swallowing in which voluntary (cortical) control of swallowing plays the largest role are the pre-oral and oral phases of swallowing: the functional components of oral feeding include food-getting, masticators efficiency, the formation of a swallow-safe bolus, and the initiation of swallowing. Patterns of oral dysfunction include tongue thrust, hyperactive and hypoactive gag reflexes, oral hypersensitivity, and prolonged, exaggerated, bite reflexes. Inadequate function of cheek and lip musculature may prevent formation of a normal oral (lip) seal, lead to food/liquid loss, and inhibit distal propulsion of an organized bolus: this affects indirectly also the pharyngeal stage of swallowing (4).

For safe swallowing to occur, oral motor function, swallowing, and respiration must be thoroughly controlled and coordinated: the impaired ability to plan and coordinate swallowing with ventilation is frequently present in CP children (5). This can lead to dysfunction of pharyngeal stage of swallowing with penetration or aspiration of food into the airways. The presence of rattling, gurgling respirations, cough with voice changes, gag, colour changes, or delayed swallow after the child has taken

[*]Correspondence: Annamaria Salghetti
Annamaria Salghetti, via Costa Alta 37 31015 Conegliano, Italy
E-mail: annamaria.salghetti@cn.lnf.it

several sucks of liquids is highly suggestive of aspiration or penetration (4-6). The clinical consequence of abnormality in the oral preparation and oral stage of swallowing is malnutrition, while the clinical consequence of aspiration is recurrent chest infections (7-8). Mildly affected children may grow satisfactorily during early childhood, which is often accomplished through more frequent feeding, prolonged mealtimes, and caloric boosting of meals. Weight-for-age does decline through childhood, but because these children are also shorter in stature they remain proportional in their weight-for-height (7).

Moderately affected children are markedly growth retarded by 3 years of age and show the characteristic decline through the adolescent years. Severely affected children show the most rapid weight decline with age, and also the characteristic growth failure in infancy (7). Clinical consequences of aspiration depend mainly on the age of the child and on the extent of aspiration itself. In young infants, aspiration is more likely to occur with apnea and/or bradycardia. In older children, it will generally be presented with cough, choking, chronic noisy breathing (congestion), recurrent wheezing (asthma, tracheomalacia), chronic/recurrent pneumonia, bronchitis and atelectasias. Breathing problems and malnutrition reduce expected life duration in CP (8). It's important to detect and treat dysphagia in order to improve the life expectancy and quality of life in these children.

2. Clinical and instrumental evaluation

There are several important features in the process of feeding that need consideration: voluntary control of oral-motor area, primitive reflexes, head and trunk control, patterns of oral dysfunction and coordination with breathing (9). Here, we will discuss these points briefly.

The functional components of oral feeding include food-getting, masticators efficiency, the formation of a swallow-safe bolus, and the initiation of swallow. The anatomical components of the oral preparatory phase involve the oral cavity, the muscles of mastication, and the musculature of the face. A disruption in the anatomical structures or physiologic function of the feeding system can severely inhibit the individual's ability to feed safely and to deliver a sufficient quantity of food to the gastrointestinal tract to be nutritionally supportive. Children with severe oral-motor impairment take 2-12 times longer to manipulate and swallow a standard amount of pureed food and 1-15 times longer to chew and swallow solid food than children of the

same weight (10). Thus, mechanical inefficiency in feeding accounts in part for the malnutrition observed in children with CP and may contribute to the low growth potential of these children.

Primitive reflexes of the normal infant such as suckle-swallow, rooting, gagging and biting are essential for infant survival and are a part of normal development. However, the prolonged persistence of these and other primitive reflexes such as the asymmetrical tonic neck reflex (ATNR), often observed in patients with developmental delay and CP can interfere with the patient's feeding skills. Abnormal responses such as bite reflex, suckle-swallow reflex, lack of tongue lateralization, instability of lower jaw and phasic biting, can severely limit the individual's ability to masticate, position, and swallow a food bolus safely (11).

A proper body position for feeding is essential to promote normal swallowing. Ideal feeding requires that a person is placed in a seated position with the buttocks well back in the chair, knees bent, and trunk and head in the midline. The neck should be in slight flexion. This position facilitates normal head and oral motor activity during feeding and maintains proper breathing patterns. It also provides good alignment of the upper alimentary tract and minimizes occurrence of the gag reflex. The degree of neck flexion/extension can influence neurologically impaired patients as they feed: neck extension can inhibit muscular movements of swallowing and can align the airway to facilitate aspiration (12). A patient who has an extensor thrust "reflex" can go into full body extension as a result of the hyperextension of the neck. This full-body extension interferes with positioning and subsequent feeding attempts, this often requires the patient to be restrained in a seat for feeding with trunk, neck and head supports (13).

A person must be able to control head and mouth movement independently from the rest of the body to be successful as a self-feeder: populations of patients with feeding disorders are naturally divided into those who can feed themselves and those who cannot (dependent feeders) (14).

Patterns of oral dysfunction include tongue thrust, hyperactive and hypoactive gag reflexes, oral hypersensitivity, and prolonged, exaggerated bite reflexes. Inadequate function of cheek and lip musculature may prevent the formation of an adequate oral (lip) seal which may lead to food/liquid loss, and inhibit distal propulsion of an organized bolus (15-16).

For safe swallowing to occur, oral motor function, swallowing, and respiration must be precisely controlled and coordinated. One must sense the presence of the food, efficiently form the bolus, and then appropriately trigger the swallowing at the correct time in the respiratory cycle. Such a controlled swallowing does not occur in a coordinated way in individuals that are severely affected by neuromotor impairments in general. Frequently, these individuals have motor impairments that may interfere with efficient food processing, delays in initiating swallowing, and inefficient oropharyngeal clearance of swallowed material (12). These abnormalities may contribute to swallowing being poorly timed in the respiratory cycle and may place the individuals at risk of aspiration (17). However, the etiology of aspiration in children with cerebral palsy remains unclear: there are a number of children with reduced pharyngeal peristalsis and pharyngeal residue after swallows who do not aspirate (18). It is stated that more severe or pervasive aspirations increase the chance of acute pneumonia, but this still does not predict whether, and when, recurrent episodes of acute infection of the lower respiratory tract will lead to chronic lung damage (5). By definition an aspiration occurs when liquid or food enters the airway and travels below the level of the vocal cords towards the lungs. Penetration occurs when liquid or food enters the airway, but does not travel below the level of the vocal cords. Children with severe neurological disability have been reported as having a high prevalence of aspiration, from 68% to 70% of cases (19). There is also a phenomenon of silent aspiration, which means that foreign material entering the trachea or lungs doesn't cause an outward sign of coughing or respiratory difficulty. It was reported a 70% incidence of aspiration and a 60% incidence of silent aspiration (16).

A diagnosis of aspiration can be made according to the clinical evaluation and above all on specific diagnostic instrumental procedures (19). Clinical evaluation of aspiration/penetration is based on the auscultation over the pharynx and trachea during swallowing and on the auscultation of the lungs before and after feeding. The presence of rattling, gurgling respirations after the child has taken several sucks of the bottle is highly suggestive of recurrent aspiration (20). It is interesting that some of these children do not demonstrate a vigorous cough in response to small episodes of aspiration, which suggests that the airway defence mechanisms in these children may be inefficient (18). At the same time, cough was found to be the best predictor of

aspiration of fluids in children (6). When cough is present with voice changes, gag, colour changes, or delayed swallow, the risk of aspiration is increased and clinicians should be aware that the child could be aspirating fluids (21). Detecting aspiration of solids may be more difficult. Pulse oximetry can also tell us something about aspiration, but it is not sensitive and specific enough (17).

Since clinical evaluation may fail to detect penetration or aspiration, particularly in the case of "silent aspiration", and is not able to assess accurately the pharyngeal and esophageal stages of swallowing, videofluoroscopy (VFS) is commonly recommended for evaluating the pharyngeal-esophageal stages of swallowing (19). Clinical detection of solid aspiration is not as accurate as the VFS, which allows us to detect the altered phase of swallowing and quantify the risk for aspiration of food in different consistency (6). Videofluoroscopy is used to identify children in whom oral feeding may be contraindicated (especially patients, with "silent aspiration") and to determine which bolus characteristics of food are swallow-safe (size, consistency) (18). Videofluoroscopy does not, however, provide quantitative data on the function of oral and pharnyngeal structures involved in deglutition. Pharyngeal manometry remains the best method for evaluating pharyngeal and esophageal motor function. Manometry has been mainly used to investigate esophageal function in children with gastroesophageal reflux and other esophageal motor disorders. Ultrasonography represents a new diagnostic technique for the evaluation of swallowing disorders. This is a noninvasive test that allows visualization of the motion of structures in the oral cavity, such as the tongue and floor of the mouth, during feeding and deglutition (10).

In addition to the problem of dysfunctional swallowing, many neurologically impaired children suffer from an associated dysfunction involving the gastroesophageal junction, known as gastroesophageal reflux (GER). This may be defined as dysfunction of the distal esophagus leading to frequent return of stomach contents into the esophagus. In children with central nervous system disease, the incidence of GER has been reported to be as high as 75% (22). The mechanism accounting for this phenomenon is unknown, although several factors may be involved, including habitual aerophagia, frequent recumbent positioning, diaphragmatic distortion secondary to kyphosis and scoliosis, and the effect of brain injury on function of the lower esophageal sphincter. The child with impaired

swallowing has a poorly protected airway and, consequently, an episode of acid reflux may result in severe pulmonary conditions such as bronchospasm, pneumonia, or apnea (8). Reflux frequently occurs shortly after feeding, but episodes have been observed up to several hours after a meal. Symptoms that suggest the presence of GER include a history of regurgitation, vomiting, night-time coughing, and vomits on the pillow in the morning, or ruminating. Esophagitis secondary to GER may present with hematemesis, refusal of food, and unexplained irritability (23).

3. Treatment

Therapy of gastroesophageal reflux is somewhat controversial. Simple measures such as positioning the child supine after feeding with the head of the bed elevated, use of thickened feedings, are often all that is needed. In infants who do not respond to these measures or who are thought to be at high risk for life-threatening events such as apnea or pneumonia, pharmacologic therapy may be useful. Surgical intervention should be reserved for the more severe cases in patients for whom aggressive medical management has failed.

In clinical practise dysphagia is often underestimated. Since the problem is complex and involves different abilities, its evaluation must be multidisciplinary. However the recognition of dysphagia signs is not easy, and only dedicated professionals can make a complete evaluation. Nevertheless, it is important for physicians to be able to detect the subpopulation of CP children that has higher risk of dysphagia. Measures of weight for height, a change in growth trajectory, skin-fold thickness, blood indices of poor nutrition, recurrent chest infections, presence of dysphagia signs, like rattling, gurgling respirations, cough with voice changes, are all quantifiable means of setting thresholds for action (4-12). If the child presents these symptoms, it is important to make more accurate clinical and/or instrumental evaluations. Experienced professionals should use their uncertainty about the presence or absence of penetration or aspiration as an indicator that further diagnostic tests are required, specifically VFS. However, swallow study with VFS provides the basis for more objective planning only if combined with careful clinical evaluation.

How aggressive therapy must be largely dependent upon the severity of the symptoms and the results of instrumental evaluation. Despite the prolonged mealtime, mildly affected children eat softer food textures than normal children, such as muffins, yogurts or bananas rather then sandwiches, apples or freshly cut vegetables. In terms of treatment it's suggested an increase in the frequency and in the caloric density of meals so as not to prolong mealtimes (24-25). In more affected children, therapy may focus on the best positioning for feeding, special utensils to facilitate feeding, examining what food textures the child can tolerate and the easiest meals for drinking (26). If the child still presents severe dysphagia signs in spite of these precautions, a temporary cessation of oral feedings may be necessary. These children have risk with routine feeding, even when it is done by capable therapists, and despite the fact that most have been in intensive swallow therapy programs. These patients also pose the greatest problems in management and raise important issues about the liability faced by therapists during the feeding. In these cases gastrostomy feeding is highly recommended. In this instance, nasogastric feedings as a temporary measure can be effective in controlling symptoms (8).

Severely affected children are diagnosed in infancy due to their inability to coordinate sucking and breathing during feeding. Children show difficulties during weaning by being unable to make the transition to eating solids and drinking liquids from a cup. Inability to gain weight and the deterioration to low weight-for-age percentiles during the first year of life are characteristic of this group of children; in these cases early and aggressive treatment to prevent growth failure is widely accepted. Children usually undergo a short period of nasogastric feeding to reinstate the growth channel shown at birth. If weight deterioration continues after a period of nasogastric feeding, gastrostomy feeding is recommended.

In general, the lack of evidence-based guidelines, the most accepted pragmatic argument has been that any clinically demonstrated aspiration may be dangerous, and if traditional techniques such as postural management and texture modification fail to eliminate it, gastrostomy should be implemented to protect the lungs and to improve weight gain. Set against this view is the fact that non-oral feeding is often perceived as a failure, not only by parents but also by many professionals involved in the care of children with disabilities. The reasons for this are deep-rooted and complex. The ability to eat is seen as a fundamental human right. Gastrostomy feeding does not preclude oral feeding as long as it is judged to be safe for the child; both of them should be coordinated carefully. Oral feeding should precede gastrostomy feeding so that the child can ingest food when he/she is hungry.

Gastrostomy feeding may relieve much of the pressure for "getting enough food into the child." Oral feeding may then focus on the pleasurable, social aspects of the meal, while gastrostomy feeding provides necessary calories for growth. Though gastrostomies are not without risk, the benefits of adequate nutrition usually outweigh the risks of complications. Because eating is so strongly embedded in our social functioning, caregivers often desire to feed their child; it is one of the few socially meaningful interactions with severely affected children. For parents, a gastrostomy might be seen as another step in hospitalization of their child. Additionally, despite the fact that parental satisfaction with gastrostomy is generally high, the procedure still has a significant morbidity and it should be performed on the basis of good justification.

4. Conclusion

Efficiency of feeding in children with eating impairments depends on accurately diagnosing the severity of such impairments. Diagnoses should be based on measures of ingestive skills that have to be done by dedicated professionals and instrumental evaluation such as Videofluoroscopy. Treatment options must then be chosen to support facilitation of eating and drinking with the aim of reducing chest infections and normalizing growth. There are some current clinical principles for feeding dysphagic CP children that have always to be remembered. In general they require good posturing during the meal and extra time to complete solid and liquid swallow; hurrying children during feeding, in attempts to complete meals in "reasonable periods of time", should be avoided. Children manage semisolid boluses more efficiently than liquid boluses. When possible, semisolid foods should be substituted for liquids in their diets. Such an approach promises to contribute substantially toward the amelioration of the quality of life of these children. Those who are severely impaired need drastic nutritional for supporting to prevent the serious malnutrition and recurrent chest infections. As the problem of dysphagia in CP children involves various aspects of their life, an interdisciplinary approach is essential for providing optimal care for these patients to manage.

References

1. Bax MC, Flodmark O, Tydeman C. Definition and classification of cerebral palsy. From syndrome toward disease. Dev Med Child Neurol Suppl 2007; 109: 39-41.

2. Hagberg B, Hagberg G, Olow I, von Wendt L. The changing panorama of cerebral palsy in Sweden. VII. Prevalence and origin in the birth year period 1987-90. Acta Paediatr 1996; 85: 954-960.

3. Gisel EG, Alphonce E. Classification of eating impairments based on eating efficiency in children with cerebral palsy. Dysphagia 1995; 10: 268-274.

4. Rogers B, Arvedson J, Buck G, Smart P, Msall M. Characteristics of dysphagia in children with cerebral palsy. Dysphagia 1994; 9: 69-73.

5. Loughlin GM. Respiratory consequences of dysfunctional swallowing and aspiration. Dysphagia 1989; 3: 126-130.

6. DeMatteo C, Matovich D, Hjartarson A. Comparison of clinical and videofluoroscopic evaluation of children with feeding and swallowing difficulties. Dev Med Child Neurol 2005; 47: 149-157.

7. Gisel EG, Patrick J. Identification of children with cerebral palsy unable to maintain a normal nutritional state. Lancet 1988; 1: 283-286.

8. Cass H, Wallis C, Ryan M, Reilly S, McHugh K. Assessing pulmonary consequences of dysphagia in children with neurological disabilities: when to intervene? Dev Med Child Neurol 2005; 47: 347-352.

9. Kenny DJ, Koheil RM, Greenberg J, et al. Development of a multidisciplinary feeding profile for children who are dependent feeders. Dysphagia 1989; 4: 16-28.

10. Casas MJ, McPherson KA, Kenny DJ. Durational aspects of oral swallow in neurologically normal children and children with cerebral palsy: an ultrasound investigation. Dysphagia 1995; 10: 155-159.

11. Selley WG, Parrott LC, Lethbridge PC, et al. Objective measures of dysphagia complexity in children related to suckle feeding histories, gestational ages, and classification of their cerebral palsy. Dysphagia 2001; 16: 200-207.

12. Tuchman DN. Cough, choke, sputter: the evaluation of the child with dysfunctional swallowing. Dysphagia 1989; 3: 111-116.

13. Waterman ET, Koltai PJ, Downey JC, Cacace AT. Swallowing disorders in a population of children with cerebral palsy. Int J Pediatr Otorhinolaryngol 1992; 24: 63-71.

14. Gisel EG. Oral-motor skills following sensorimotor intervention in the moderately eating-impaired child with cerebral palsy. Dysphagia 1994; 9: 180-192.

15. Rempel G, Moussavi Z. The effect of viscosity on the breath-swallow pattern of young people with cerebral palsy. Dysphagia 2005; 20: 108-112.

16. Griggs CA, Jones PM, Lee RE. Videofluoroscopic investigation of feeding disorders of children with multiple handicap. Dev Med Child Neurol 1989; 31: 303-308.

17. Morgan AT, Omahoney R, Francis H. The use of pulse oximetry as a screening assessment for paediatric neurogenic dysphagia. Dev Neurorehabil 2008; 11: 25-38.

18. Mirrett PL, Riski JE, Glascott J, Johnson V. Videofluoroscopic assessment of dysphagia in children with severe spastic cerebral palsy. Dysphagia 1994; 9: 174-179.

19. Sorin R, Somers S, Austin W, Bester S. The influence of videofluoroscopy on the management of the dysphagic patient. Dysphagia 1988; 2: 127-135.

20. Judd PL, Kenny DJ, Koheil R, Milner M, Moran R. The multidisciplinary feeding profile: a statistically based protocol for assessment of dependent feeders. Dysphagia 1989; 4: 29-34.

21. Selley WG, Parrott LC, Lethbridge PC, et al. Objective measures of dysphagia complexity in children related to suckle feeding histories, gestational ages, and classification of their cerebral palsy. Dysphagia 2001; 16: 200-207.

22. Arvedson J, Rogers B, Buck G, Smart P, Msall M. Silent aspiration prominent in children with dysphagia. Int J Pediatr Otorhinolaryngol 1994; 28: 173-181.

23. Lefton-Greif MA, Carroll JL, Loughlin GM. Long-term follow-up of oropharyngeal dysphagia in children without apparent risk factors. Pediatr Pulmonol 2006; 41: 1040-1048.

24. Gisel EG. Effect of oral sensorimotor treatment on measures of growth and efficiency of eating in the moderately eating-impaired child with cerebral palsy. Dysphagia 1996; 11: 48-58.

25. Gisel EG, Applegate-Ferrante T, Benson J, Bosma JF. Oral-motor skills following sensorimotor therapy in two groups of moderately dysphagic children with cerebral palsy: aspiration vs nonaspiration. Dysphagia 1996; 11: 59-71.

26. Gisel EG, Applegate-Ferrante T, Benson JE, Bosma JF. Effect of oral sensorimotor treatment on measures of growth, eating efficiency and aspiration in the dysphagic child with cerebral palsy. Dev Med Child Neurol 1995; 37: 528-543.