Residência Pediátrica
Publicação Oficial da Sociedade Brasileira de Pediatria
ISSN-Online: 2236-6814

Submitted on: 12/05/2019
Approved on: 04/13/2020

POINT OF VIEW

Respiratory failure in pediatric patients with cerebral palsy: how can we prevent it?

Jáder Pereira Almeida[1], Sylvio Gilberto Andrade Ávilla[2], Paulo Ramos David João[1]

Keywords:
Pediatrics,
Cerebral Palsy,
Primary Prevention,
Respiratory Insufficiency.

Abstract

Pediatric patients with cerebral palsy have a higher risk of respiratory failure due to their anatomical and physiological particularities, requiring the adoption of preventive treatment to improve their quality of life. Topics such as working with multidisciplinary team, feeding methods and respiratory secretion mobilization strategies are discussed in this article. In addition, treatment of sialorrhea, gastroesophageal reflux, bronchiectasis, malnutrition and laryngotracheal separation surgery are addressed, with the aim of reducing respiratory morbidity in these patients.

[1] Pequeno Príncipe Hospital, Pediatric Intensive Care Unit - Curitiba - Paraná - Brazil.
[2] Pequeno Príncipe Hospital, Department of Pediatric Surgery - Curitiba - Paraná - Brazil.

Correspondence to:
Jáder Pereira Almeida.
Pequeno Príncipe Hospital. Rua Desembargador Motta, 1070 - Água Verde, Curitiba/PR, 80250-060. E-mail: jader.pa@hotmail.com

DOI: 10.25060/residpediatr-2021.v11n3-232

This work is licensed under a Creative Commons Attribution 4.0 International License.

Cerebral palsy (CP) is a group of disorders that affect the individual's motricity in varying degrees of severity, and may be accompanied by disorders of sensitivity, cognition, perception or communication[1]. The most common subtype is the spastic form, corresponding to 80% of cases, characterized by high muscle tone and dysfunction of body mobility. Among the main risk factors, we can highlight prematurity, gestational infections and tocotraumatism[2].

Despite the technological advances of the last decades, part of the children with CP with severe dysfunction (Grades IV and V by the *Gross Motor Function score Classication System)* have a low quality of life, with frequent visits to the pediatric emergency room, and respiratory compromise is the main cause of morbidity and mortality[2-4].

Some anatomical and physiological characteristics may explain the higher occurrence of respiratory failure in this population: (1) swallowing disorders favor the increased incidence of aspiration pneumonia; (2) kyphoscoliosis results in a reduction in the number of alveoli and chest wall compliance; (3) respiratory muscle weakness compromises tidal volume and vital capacity; and (4) ineffective cough reflex increases the risk of infections and atelectasis[1,5].

In a recent survey carried out at our institution, it was observed that approximately 20% of admissions to the general and surgical intensive care unit (ICU) are due to respiratory failure, with one third of those analyzed having CP. Other authors have identified similar results[6]. Therefore, we can see the importance of the topic in the context of critically ill patients.

When children with CP are admitted to the ICU for respiratory failure, the first line of treatment is the use of non-invasive mechanical ventilation. This support modality can be used in patients with or without blood gas changes, with the aim of making the respiratory cycle more effective. However, despite the good efficacy in the general population, patients with CP are at higher risk of therapeutic failure due to reduced mucociliary *clearance* , higher prevalence of upper airway obstruction and greater difficulty in adjusting the ventilator interface to the patient[7,8]. Consequently, they are more frequently submitted to orotracheal intubation and its complications.

Once the acute condition is resolved, they also have a higher risk of failure to wean from mechanical ventilation. Hsia et al. (2012)[9] identified this problem in their retrospective study evaluating 139 children dependent on mechanical ventilation, and a large part of the sample had CP requiring ventilatory support for more than 5 years. It is noteworthy that prolonged mechanical ventilation is also a risk factor for the occurrence of pneumonia.

Thus, we can see that despite the technological apparatus, children with CP with respiratory failure are at greater risk of therapeutic failure, and efforts should be made in preventive measures. Based on the experience of our institution and on data from the medical literature, we address some measures that can improve the quality of life of these patients.

- **Working with a multidisciplinary team**: it is necessary to involve some specialists in the follow-up of these patients due to the complexity of the condition, not only for the management of risk factors that result in respiratory failure, but also for psychosocial issues involving parents and guardians. In our unit, the team is made up of a pediatrician, pediatric neurologist, pediatric pulmonologist, pediatric gastroenterologist, pediatric surgeon, otolaryngologist, physical therapist, nurse, speech therapist, nutritionist, psychologist and social worker. Professionals must work together in order to propose the best therapeutic plan to the patient.
- **Feeding methods**: CP patients may present swallowing disorders, both for solids and liquids, resulting in microaspirations, aspiration pneumonia and, consequently, respiratory failure. The suspicion must be evaluated by the swallowing specialist, and feeding and positioning techniques can be beneficial in avoiding the problem. Diagnostic confirmation is performed through videodeglutogram . When oral feeding is unfeasible or when caloric support is insufficient, a gastrostomy tube is indicated.[1] However, it is important to emphasize that alternative feeding route does not prevent tracheal aspiration of saliva. [5]
- **Treatment of sialorrhea:** Some anticonvulsants and neuroleptics can result in excessive production of saliva and bronchial secretion. Evaluation by the pediatric neurologist, in this case, may be necessary to adjust medication doses or change drugs. In addition, anticholinergic agents (eg, scopolamine) can be used to reduce saliva production, although this measure has a low level of evidence in the literature. [1] As a last resort, some authors argue that surgical ligation of the salivary ducts is a good option to reduce the number of recurrent lower respiratory tract infections. [10]
- **Effective elimination of respiratory secretions**: work with respiratory physiotherapy makes it possible to mobilize bronchial secretions through inspiratory and expiratory movements with the aid of the bag-valve-mask device. Another alternative is mechanical insufflation- deflation (IE-M) devices, helping the patient to cough in a non-invasive way. [7] It consists of performing positive pressure alternating with negative pressure with high expiratory flow in the airway, thus avoiding the deleterious effects of reduced mucociliary clearance . Siriwat et al., in their randomized clinical trial, found that IE-M is beneficial in the treatment of children with spastic cerebral palsy presenting with pneumonia and atelectasis. [11]

- **Gastroesophageal reflux treatment**: Esophageal motility disorders and abdominal muscle spasticity may justify the higher incidence of gastroesophageal reflux in patients with CP, potentiating the occurrence of aspiration pneumonia. [1] In patients with asthma, this problem is even more harmful, resulting in chronic airway inflammation and wheezing attacks . The use of rigid or flexible fibroscopy can be useful in identifying the effects of gastric contents on the respiratory mucosa and pH monitoring can confirm the diagnosis. Clinical treatment can be performed with thicker food, H2 receptor antagonists or proton pump inhibitors. When clinical measures fail, fundoplication is an effective alternative to reduce respiratory morbidity caused by reflux. [12]
- **Treatment of bronchiectasis**: Patients who experience chronic tracheal aspiration may develop bronchiectasis. Piccione et al. identified that children with severe neurological impairment and gastroesophageal reflux have a 9.4-fold ($p < 0.004$) and 3.3-fold ($p = 0.036$) greater risk of developing this problem, respectively. [13] History of chronic cough, recurrent pneumonia and/or persistence of radiographic alterations may be part of the clinical picture, and confirmation is performed by computed tomography of the chest with thin sections. Prophylactic antibiotic therapy with macrolides (eg, azithromycin every other day) may prevent exacerbations. In refractory cases, a surgical approach is indicated.
- **Malnutrition treatment:** studies show that up to 20% of CP patients may be malnourished. In situations of hemodynamic instability, increased protein catabolism and atrophy of the respiratory muscles may occur. [1] The return to food should be early, in view of its benefits. The nutrition specialist should accompany the case during hospitalization and outpatient follow-up, when possible.
- **Laryngotracheal Separation (LTS)**: in refractory cases, the evaluation of the pediatric surgeon and the indication of this procedure can be beneficial for the patient. It consists of closing the proximal segment of the trachea blindly, separating it from the distal segment that will be connected to the skin of the anterior cervical region. [14] It differs from tracheostomy in that there is no additional risk of micro-aspiration through the space between the tracheostomy tube and the trachea. Hara et al., in their study with children with neurological disease, observed a significant reduction in aspiration pneumonia after TLS. [15] In our unit, the procedure is performed in selected patients, with good results, with a low complication rate.

REFERENCES

1. Boel L, Pernet K, Toussaint M, Ides K, Glenn L, Haan J, et al. Respiratory morbidity in children with cerebral palsy: an overview. Dev Med Child Neurol. 2019 Jun;61(6):646-53.
2. Ray S, Chatterjee S, Ruiz-Lozano P. Pediatric cerebral palsy life expectancy: has survival improved over time? Pediatr Ther. 2013;3(1):146.
3. Vilela LR, Brízida LVO, Pádua MF, Silva BM, Ferreira LS. Perfil de crianças e adolescentes internados por distúrbios neurológicos. Resid Pediatr. 2019;9(3):1-5.
4. Himmelmann K, Beckung E, Hagberg G, Uvebrant P. Gross and fine motor function and accompanying impairments in cerebral palsy. Dev Med Child Neurol. 2006 Jun;48(6):417-23.
5. Proesmans M, Vreys M, Huenaerts E, Haest E, Coremans S, Vermeulen F, et al. Respiratory morbidity in children with profound intellectual and multiple disability. Pediatr Pulmonol. 2015 Out;50(1):1033-8.
6. Yates K, Festa M, Gillis J, Waters K, North K. Outcome of children with neuromuscular disease admitted to paediatric intensive care. Arch Dis Child. 2004 Fev;89(2):170-5.
7. Gauld LM. Airway clearance in neuromuscular weakness. Dev Med Child Neurol. 2009 Mai;51(5):350-5.
8. Lis ARB, Duarte MCMB, Andrade LB. Noninvasive ventilation as the first choice of ventilatory support in children. Rev Bras Ter Intensiva. 2019 Out;31(3):333-9.
9. Hsia SH, Lin JJ, Huang IA, Wu CT. Outcome of long-term mechanical ventilation support in children. Pediatr Neonatol. 2012 Out;53(3):304-8.
10. Manrique D, Sato J. Salivary gland surgery for control of chronic pulmonary aspiration in children with cerebral palsy. Int J Pediatr Otorhinolaryngol. 2009 Set;73(9):1192-4.
11. Siriwat R, Deerojanawong J, Sritippayawan S, Hantragool S, Cheanprapai P. Mechanical insufflation-exsufflation versus conventional chest physiotherapy in children with cerebral palsy. Respir Care. 2018 Fev;63(2):187-93.
12. Sload RL, Brigger MT. Surgery for reflux induced airway disease: a systematic review. Int J Pediatr Otorhinolaryngol. 2014 Ago;78(8):1211-15.
13. Piccione JC, McPhail GL, Fenchel MC, Brody AS, Boesch RP. Bronchiectasis in chronic pulmonary aspiration: risk factors and clinical implications. Pediatr Pulmonol. 2012 mai;47(5):447-52.
14. Antunes LA, Talini C, Carvalho BC, Guerra JP, Aristides ES, Oliveira DE, et al. Laryngotracheal separation in pediatric patients: 13-year experience in a reference service. Einstein (São Paulo). 2019;17(3):eAO4467.
15. Hara H, Hori T, Sugahara K, Ikeda T, Kajimoto M, Yamashita H. Effectiveness of laryngotracheal separation in neurologically impaired pediatric patients. Acta Otolaryngol. 2014 Jun;134(6):626-30.