Respiratory morbidity in children with cerebral palsy: an overview

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Respiratory problems have a significant impact on morbidity and mortality in patients with cerebral palsy (CP). In particular, recurrent aspiration, impaired airway clearance, spinal and thoracic deformity, impaired lung function, poor nutritional status, and recurrent respiratory infections negatively affect respiratory status. Bronchopulmonary dysplasia may contribute to pulmonary problems, but asthma is not more common in CP than in the general population. We discuss treatment options for each of these factors. Multiple coexisting and interacting factors that influence the respiratory status of patients with CP should be recognized and effectively addressed to reduce respiratory morbidity and mortality.

Cerebral palsy (CP) describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain.[1] The motor disorders of CP are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour; by epilepsy and by secondary musculoskeletal problems. CP is a common cause of physical disability in childhood, with reported prevalences ranging from 1 to 4 per 1000 live births and a higher prevalence in males than in females.[2] Patients with CP are a very heterogeneous group: type and severity of symptoms, degree of functioning, and quality of life are highly variable. Different subtypes can be distinguished and should be characterized according to the nature and typology of the motor abnormalities, the functional motor abilities, accompanying impairments, anatomical and neuroimaging findings, and causation and timing.[1]

Life expectancy for people with CP is shorter than for the general population and has changed little over time,[3] although a recent study by Brooks et al.[4] did show significant improvements over the past three decades in California. Survival is related to the severity of disability. Life expectancy decreases with increasing severity of motor and cognitive impairment; therefore life expectancy is lower for individuals in higher Gross Motor Function Classification System (GMFCS) levels.[3,5] Respiratory diseases are the most frequent reported cause of morbidity and mortality in CP, hence, by ameliorating respiratory status, quality of life and life expectancy might be augmented.[3,5–10] Here, we review factors that influence the respiratory status of patients with CP and describe treatment options.

As members of a multidisciplinary team caring for patients with CP in a tertiary hospital and a rehabilitation centre, we regularly encounter respiratory problems in these patients. Based on our experience and a non-systematic literature search performed in PubMed, Embase, the Cochrane Database of Systematic Reviews, and the Cochrane Controlled Trials Register, we provide an overview on respiratory morbidity in CP. The present paper does not cover sleep disorders in CP, nor does it go into detail about specific treatments. The sections of this review are organized according to the clinical management of a child with CP and chronic or recurrent respiratory symptoms.

Our centres have a specific clinic for the management of respiratory disorders in children with CP. During a first visit, the following items are routinely assessed, which follows the outline of this review: recurrent aspiration, nutritional status, airway clearance, upper airway ventilation, and deformity of the spine and chest wall. All these factors influence lung function and the risk of recurrent respiratory infections, and, ultimately, respiratory failure. Other factors, such as asthma and bronchopulmonary dysplasia, are also discussed.

Abnormal muscle tone, whether increased or decreased, plays an extremely important role, although tone management is not in the scope of this paper. Non-motor disorders may also be important and justify the need for a multidisciplinary approach.

Patients with CP are prone to pulmonary infections, but respiratory problems are not always easily recognized and diagnosed. Sometimes, this may be due to: (1) indirect communication with the patient via a parent or carer, (2)

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the fact that investigations are difficult to perform, and (3) that initial symptoms may be very subtle. This may cause delay in diagnosis and treatment, increasing the risk for complications in this vulnerable population, although experienced parents usually do notice subtle signs early.[6,7,11]

Respiratory status is influenced by a variety of factors that mostly coexist and interact. These factors and the possible treatment options are summarized in Table SI (online supporting information). Recurrent aspiration, impaired airway clearance, and deformity of the spine and/or chest wall are the most important causes of both acute respiratory tract infections as well as chronic lower airway inflammation, eventually resulting in bronchiectasis. Bronchiectasis impairs clearance of sputa and induces recurrent chest infections.[11–14]

RECURRENT ASPIRATION

Aspiration can result in aspiration pneumonia, an acute infectious event, but it is an important cause of chronic airway inflammation as well. Recurrent aspiration can be caused by oropharyngeal motor dysfunction and/or gastro-oesophageal reflux.

Oropharyngeal motor dysfunction

Swallowing is a complex process, requiring well-coordinated action of muscles in the mouth, pharynx, larynx, oesophagus, and diaphragm. In CP, poor coordination accounts for the occurrence of oropharyngeal motor dysfunction and sequential direct aspiration. In this context, it is important to mention Worster-Drought syndrome, which is classified as CP and associated with severe bulbar dysfunction and a high incidence of eating difficulties.[15] van den Engel-Hoek et al.[16] compared videofluoroscopic findings in patients with CP and neuromuscular diseases. Children with CP demonstrated dysphagia in one or all phases of swallowing, while in neuromuscular disorders, muscle weakness mainly resulted in pharyngeal residue after swallowing. The authors concluded that the underlying swallowing problem in neuromuscular disorder is muscle weakness, whereas in CP it is more complex and relates to abnormal control of swallowing.[16] Direct aspiration can happen at any stage of the swallowing process and is more frequent with ingestion of liquids. Aspiration is reported to occur in 26% to over 70% of children and adults with CP.[11,14,17] When swallowing difficulties are suspected, clinical assessment including history taking and observation of eating and drinking by a speech and language therapist are recommended as the first-line investigation.[18] A videofluoroscopic swallowing study is useful in the diagnosis of dysphagia and aspiration, but should, according to the UK National Institute for Health and Care Excellence (NICE) guideline, only be considered by a specialist multidisciplinary team in specific cases, including uncertainty about the safety of swallowing after specialist clinical assessment, recurrent chest infections without clinical signs of aspiration, deterioration in eating, drinking and swallowing with increasing age, uncertainty about the impact of modifying food textures, and in case parents or carers need better understanding of the swallowing to help with decision-making.[19] Aspiration can occur while eating, from unsafe swallowing of liquids or solids, as well as between meals by aspiration of saliva and secretions of the upper respiratory tract.[6,11,14,17] In case of aspiration while eating, positioning, feeding techniques, and equipment need to be optimized. Strategies for developing oral motor skills should be practised, and the texture of fluids and food modified according to individual needs. If these measures are insufficient, tube feeding, via either a nasogastric tube or gastrostomy, should be provided.[6,11,14,17] Sullivan et al.[14] showed in a prospective study that tube feeding by gastrostomy leads to a decrease in chest infections, direct aspiration, and choking episodes. On the other hand, in their retrospective chart review Proesmans et al.[20] do not report a decrease in respiratory infections in patients with a gastrostomy. This may be attributed to the fact that patients with more severe disabilities more often need a gastrostomy and that a gastrostomy does not prevent gastro-oesophageal reflux, nor does it prevent aspiration of saliva or upper airway secretions.[20]

What this paper adds

- Respiratory problems are a significant cause of morbidity in patients with cerebral palsy (CP).
- Respiratory status in patients with CP is influenced by recurrent aspiration and impaired airway clearance.
- Spinal and thoracic deformity, impaired lung function, poor nutrition, and respiratory infections also negatively affect respiratory status.
- These factors should all be addressed to reduce respiratory problems in patients with CP.

Aspiration of saliva

If saliva aspiration occurs, currently used medication should be reviewed, because some anti-epileptic drugs (e.g. clobazam, clonazepam) and neuroleptics induce drooling. Gastro-oesophageal reflux should be treated (see below) as chemical irritation increases saliva production. Treatment options to reduce saliva production include anticholinergic agents (e.g. glycopyrronium bromide [enteral], scopolamine [transdermal]), botulinum neurotoxin A (BoNT-A) injection in the salivary glands, or surgery (duct ligation, gland removal).[16,17] These treatments are effective in reducing the amount of saliva and drooling, but data concerning the effect on respiratory status are missing for anticholinergic drugs[21,22] and rather limited for BoNT-A injection and surgery. BoNT-A injection in the salivary glands has been said to be effective in reducing respiratory problems in patients with neurological impairment, but further evidence is needed.[23,24] It should also be taken into account that BoNT-A injection in the salivary glands can be associated with mild and transient oral motor problems, especially in patients with CP, those with mild drooling before treatment, or if a high dose of BoNT-A is used.[25] Data about surgical treatments are conflicting. Some authors reported that surgical saliva reduction was effective in

reducing respiratory infections,[26,27] but others found no significant reduction in respiratory infections.[28] The usefulness of oral motor therapy has not been systematically studied in children with CP. Suctioning of saliva through a suction catheter during daytime can be used as a supportive measure in patients with sialorrhea, although frequent suctioning might drive saliva production.

Gastro-oesophageal reflux

Gastro-oesophageal reflux is common in children with CP (incidence 32%–75%). Possible reasons are increased intra-abdominal pressure from spasticity of abdominal muscles and/or poor coordination between oesophageal movement, and sphincter activity. The refluxed gastric content may be aspirated because of oropharyngeal motor dysfunction. This mechanism is termed indirect aspiration.[11,14,17] In some centres, gastroscopy and/or bronchoscopy are used to assess gastro-oesophageal reflux because flexible endoscopy can demonstrate the effects of reflux on the mucosae. Bronchoscopy with bronchoalveolar lavage and determination of the lipid laden macrophages index is a sensitive way to prove pulmonary aspiration, as an increased presence of lipid laden macrophages in the lower airways suggests aspiration of food, gastro-oesophageal reflux may be addressed using feed thickening, medication (histamine H_2 antagonist, proton pump inhibitor), or by fundoplication.[11,14,17] Medical treatment of gastro-oesophageal reflux improves respiratory symptoms in children and proton pump inhibitors are more efficacious than histamine H_2 antagonists.[29] Despite treatment with proton pump inhibitors, gastric juices still cause a significant inflammatory reaction when bronchial epithelial cells are exposed to it.[30] Further studies are needed to clarify the effect of medical treatment of gastro-oesophageal reflux on respiratory morbidity in patients with CP. In one study, children with CP who underwent percutaneous endoscopic gastrostomy placement alone were compared with children with CP who underwent both percutaneous endoscopic gastrostomy and laparoscopic fundoplication at the same time regarding their respiratory outcome.[14] No significant difference in the use of antibiotics or hospitalization for respiratory infections was found. Nevertheless, in both children and adults with gastro-oesophageal reflux-related airway disease but without neurological impairment, fundoplication has been shown to be effective.[31–33]

Direct and indirect aspiration may coexist and both can occur without obvious coughing or choking, known as 'silent aspiration'. This could be due to diminished sensitivity of cough receptors by chronic aspiration.[11,17,19]

Poor nutritional status

It is reported that about 20% of children and young people with CP are undernourished.[18] Patients with CP are at risk of malnutrition because of feeding problems, gastro-oesophageal reflux, and additional energy expenditure. In a catabolic state, respiratory muscles atrophy, lung function declines, bacterial colonization of the airways increases, and resistance to infection decreases. To prevent poor nutritional status, a speech therapist should assess the role of oropharyngeal motor dysfunction, gastro-oesophageal reflux should be treated appropriately, and a dietician can optimize calorie intake by adding supplements. If all these measures are unsatisfactory tube feeding may be necessary.[11,14]

Impaired airway clearance

Airway clearance by cough is the most important factor in the prevention of pulmonary infections, but cough may be ineffective in CP because of respiratory muscle weakness and/or poor coordination. Coughing is an important but complex reflex which protects the lower airways by removal of mucus and other harmful substances. When cough is weak, as in children with CP, secretions are inadequately cleared, which predisposes to infection and atelectasis.

To ameliorate coughing, positioning is very important, as slumped sitting increases respiratory effort and makes it harder to cough.[6]

Cough can be stimulated by augmenting inspiration, expiration, or both.[34] Augmenting inspiration consists of air stacking by glossopharyngeal breathing or by using a resuscitator bag or ventilator. However, this requires adequate control of the upper airway by the patient, which is not feasible in patients with severe neurodevelopmental delay. When augmenting expiration, an upward thrust is applied to the epigastrium or anterior chest wall by a carer while the patient coughs. A mechanical insufflator-exsufflator 'imitates' a cough: it generates a positive pressure in the airway to air stack followed by a switch to a negative pressure causing a high expiratory flow.[34]

Because of chronic infection, airway clearance may be impaired. Physiotherapy techniques to promote airway clearance are necessary in children with CP. Besides classic drainage, positive expiratory pressure and intrapulmonary percussion ventilation systems can be used by the physiotherapist. By using a positive expiratory pressure device, the patient exhales against a constant, adjustable resistance. Because of this resistance, the intrabronchial pressure increases and airways are prevented from collapsing, which facilitates clearance of mucus. Intrapulmonary percussion ventilation causes vibration of air in the lungs, which loosens airway secretions and facilitates their drainage. Although studies concerning physiotherapy in CP specifically are lacking, respiratory physiotherapy may be necessary daily, but is especially important during respiratory infections.[6,11] This is, for instance, also illustrated in a recent study where a community respiratory physiotherapy service was installed for children with disabilities with an acute infection. This resulted in a reduction of hospital admissions and bed days for these children.[35] A relatively new device, the Free-Aspire (Medical Products Research, Srl, Legnano, Italy) is an electro-medical machine for removing secretions. The device utilizes Vakűm technology: during expiration the airflow is accelerated by the

Venturi effect inside a special connector. The acceleration is activated only during the expiratory phase, is proportional to the flow of air on spontaneous breathing, and does not require any cooperation. The secretions slide along the layer of liquid lining the bronchial epithelium until they reach the glottis from where they are either expectorated or swallowed. No negative pressure is generated inside the airways, avoiding a possible risk of collapse. In a small pilot study in patients with CP, the use of this machine was associated with less use of antibiotics and hospitalizations.[36]

Upper airway obstruction and hypoventilation

Although it is not the main focus of this paper, upper airway obstruction, both awake and during sleep, with resulting hypoventilation, is an important contributor to respiratory morbidity and decreased quality of life in individuals with CP. The presence of severe CP and/or epilepsy can further aggravate this risk.[37] The clinician must be aware of symptoms of upper airway obstruction both during daytime and during sleep. Investigations can include polysomnography, imaging of the upper airway, and awake and drug-induced endoscopy of the airway to identify the severity and the cause of the upper airway obstruction. Management can include surgery, non-invasive ventilation, or tracheostomy in very severe cases.[6] The requirement for tracheostomy might increase with age.[38] Positioning and tone management are also important treatment strategies that can normalize the breathing pattern in individual patients. Although upper airway obstruction, both during sleep and wakefulness, is prevalent in patients with CP with a clear association with lower quality of life, the link with recurrent respiratory infection is not well investigated. The management options are diverse ranging from non-invasive treatments to very invasive options including tracheostomy, where ethical issues arise.[39] In our opinion, it is important to identify the exact sites of obstruction, for instance by imaging or endoscopy, to select the most appropriate treatment.[40] For severe multilevel obstruction, non-invasive ventilation is the primary option, at least in our centre. Although the burden of care needs to be assessed, it is our opinion that establishing a normal breathing pattern in these patients results in a better quality of life and, possibly, in less hospital admissions. This requires a multidisciplinary unit, experience in non-invasive ventilation, and a teaching programme so that non-invasive ventilation can be used at home by the parents[41] or in residential treatment centres.

Deformity of the spine and chest wall

During spinal growth, different events occur simultaneously and are well synchronized; if this growth process is interfered with by unequal muscle tone, a spinal deformity occurs.[42] Early-onset spinal and thoracic cage deformity interferes with lung development, reflected in a reduced number of alveoli.[42] Curvature, rotation, and shortening of the spine result in thoracic cage deformity. A deformed thoracic cage causes reduced lung volume, decreased lung compliance, increased stiffness of the chest wall, reduced force of the respiratory muscles, and increased mechanical dysfunction of the diaphragm. Lung disease in these patients is restrictive in nature[43] and unequal lung expansion causes ventilation-perfusion mismatch.[11] The increased work of breathing in combination with the above-mentioned changes predispose to respiratory failure. Alveolar hypoventilation, hypercapnia, and hypoxaemia cause pulmonary hypertension, cor pulmonale, and eventually right-sided heart failure.[43]

The general reported incidence of scoliosis in CP is 20% to 25%, but the risk increases from a very low risk in patients in GMFCS levels I and II to 50% with a more severe curve and is more likely to progress in patients in GMFCS levels IV and V. Scoliosis is progressive in nature and in these patients it may progress even after growth has ceased, which warrants early treatment.[44,45]

Conservative treatment options consist of seating arrangements and braces to provide external support to the spine. The use of braces may slow down, but does not stop, progression of scoliosis and is not always well tolerated by the patients.[11,16,44]

Surgery to correct the deformity and prevent progression should be performed early, while lung function allows, to reduce the risks of the operation and to prolong survival.[11] However, before proceeding to surgery the risks and benefits should be discussed with a multidisciplinary team.[46] Sewell et al.[45] report a postoperative improvement in activity and participation in children with CP (GMFCS levels IV and V) and significant scoliosis. In patients with idiopathic scoliosis, surgery may improve pulmonary function, but these data are lacking concerning patients with CP, although this is one of the goals for this surgery.[43,47] Another purpose for scoliosis surgery in patients with CP is to decrease the incidence of pneumonia; however, there is little evidence that this is the case.[47] A recent systematic review concluded that there is only limited high-quality evidence for outcomes after scoliosis surgery in children with CP and that this procedure is associated with a moderately high complication rate. The intervention appears to be indicated for deformity correction, but currently there is insufficient evidence to make recommendations for this surgery as a way to also improve functional outcomes, caregiver outcomes, and quality of life.[48] Another complication of these and other orthopaedic complications is chronic pain, which is highly prevalent in children with CP in a high GMFCS level.[49] The impact of chronic pain on the respiratory system, including the effects of analgesics, remains to be studied.

Impaired lung function

Possible hypotheses for a lower pulmonary function in patients with CP are kyphoscoliosis, low muscle strength as a key mechanical factor, and reduced chest mobility; all causing restrictive lung function. Indeed, in most patients with CP lung function is lower than in typically developing

controls. Kwon and Lee compared 25 children with spastic diplegic and hemiplegic CP with 14 children with normal development and demonstrated significant differences for maximal inspiratory pressure and maximal expiratory pressure between children with and without CP.[50] Pulmonary function testing showed significant differences in forced vital capacity and forced expiratory volume in one-second between children with normal development and children with spastic diplegic CP.[50] Lampe et al.[8] measured lung vital capacity using a spirometer in 46 adults with CP (GMFCS levels I–IV) and compared these results with calculated normal values. Values of vital capacity from patients with CP were lower than the matched normative values and decreased with increasing GMFCS level.[8] The requirement for a lung function analysis is that the cognitive abilities are sufficient. This is not the case for many patients with severe CP and classic lung function measurements are not possible in these patients. The use of nocturnal capnography and oxygen saturation measurements can then be used as a proxy for respiratory muscle strength.

Motor impairment results in limited physical activity, loss of muscle mass, weakening of respiratory muscles, and compromised pulmonary function.[3,50,51] Consequently, and not surprisingly, children with CP who are able to walk, have better respiratory muscle strength and respiratory training resulting in better respiratory function (maximal inspiratory pressure, maximal expiratory pressure, and forced vital capacity).[51]

Ersöz et al.[9] demonstrated decreased chest mobility in patients with spastic CP, independent of their cognitive status, and this was more prominent in older children. In the study by Lampe et al.[8], mean values of chest expansion and vital capacity were lower than normal in adults with CP (GMFCS levels I–IV) and lower in patients with more severe motor impairment (i.e. higher GMFCS level). In patients with scoliosis, chest expansion and vital capacity have a larger deviation from normal values than in patients without scoliosis, although mean values were equal in both groups. Oxygen saturation was normal in all patients with CP despite their limited chest expansion and vital capacity. This suggests an adaptation to their reduced mobility.[8] Poor coordination, weakness of trunk muscles, and spasticity may account for the limited chest mobility in patients with CP. Resultant stiffening of the costovertebral joints and shortening of the respiratory muscles, because of shallow breathing, may worsen it. Besides these factors, children with CP only engage in limited physical activity, and since intensive physical activity promotes chest mobility, this is suggested as another possible reason for their decreased chest mobility. Reduced chest mobility interferes with normal lung function, therefore respiratory rehabilitation should be initiated early.[8,9]

Poor postural control has a negative effect on lung function, and vital capacity varies with sitting position. Anterior seat inclination extends the trunk in a more upright position, allows respiratory muscles to function more efficiently, and improves respiratory function. The reverse of the positive effect on respiratory function is that sitting on an anterior inclined seat for a long period may cause excessive lumbar lordosis and low back pain, so different sitting positions should be used alternately.[52,53] To the best of our knowledge, other forms of postural therapy have not been studied systematically.

(Recurrent) respiratory infections

All the aforementioned factors predispose to pulmonary infections. Bronchiectasis, a predisposition to recurrent lower airway infections, is mentioned as a possible consequence of recurrent aspiration,[11] but exact data are scarce. Piccione et al.[54] report bronchiectasis in 66% of children with confirmed chronic pulmonary aspiration. They identified severe neurological impairment and history of gastro-oesophageal reflux as risk factors.

Respiratory infections in patients with CP are generally thought to be common.[6,7,11] However, Proesmans et al.[20] found that the number of children with profound intellectual disability and multiple disabilities needing hospital admission for respiratory disease and the number of airway infections requiring treatment was lower than expected. Most patients did not need hospitalization, only a small subgroup had multiple admissions. On the other hand, in the study by Millman et al.[13] about 12% of children hospitalized with respiratory infections had a neurological disorder. They were significantly older (median 4y 2mo), needed a longer hospital stay, and were more likely to be admitted to the intensive care unit (36.4%).[13] Detection of pathogens decreases with increasing age among all children; however, in children with neurological disorders respiratory pathogens (respiratory syncytial virus, adenovirus, rhinovirus) are significantly less detected. Detection of bacteria was similar in all children, except for Streptococcus pneumonia, which was more common in children without underlying conditions.[13]

Adequate treatment of acute airway infections is essential to prevent complications. In children with CP, intercurrent respiratory infections should be treated with antibiotics and a prolonged course of 3 to 4 weeks may be required. Which antibiotic to use is ideally guided by the result of a (previous) sputum culture.[6]

To prevent recurrent chest infections, antibiotics (e.g. trimethoprim or azithromycin) can be used prophylactically. Azithromycin, a macrolide antibiotic, has both antibacterial and anti-inflammatory properties and is thought to reduce virulence factors. It also has few side effects, is not expensive, and has pharmacokinetic and bioavailability properties that make it appropriate as a prophylactic treatment.[6,55]

As with typically developing children, children with CP should be vaccinated per the currently used vaccination schedule. Special attention should be paid to the annual influenza vaccination.[6]

Miscellaneous factors

Asthma

Asthma is commonly diagnosed in children with CP, although there is no evidence that asthma is more common

in children with CP than their typically developing peers. When diagnosing asthma, one often needs to rely on a suggestive history and it can be difficult for parents to differentiate between wheezing and other respiratory sounds. Moreover, chronic bronchial inflammation due to chronic aspiration can also result in wheezing. Therefore, because of the risk for overtreatment, asthma therapy should be stopped if it has no effect.[6,11]

Bronchopulmonary dysplasia

Patients with CP who were born preterm may have bronchopulmonary dysplasia, which may contribute to their pulmonary problems.[11] This is most likely due to impaired lung function, because bronchiectasis is not commonly associated with bronchopulmonary dysplasia.[56] Although gram-negative bacterial colonization of the airways in the neonatal period is associated with more severe bronchopulmonary dysplasia, it is unknown if bronchopulmonary dysplasia in later life is an independent risk factor for gram-negative colonization.[57]

Dystonia and tone management

Dystonia is characterized by the presence of fluctuating hypertonia and/or involuntary postures and movements triggered by arousal, such as wakening from sleep, tiredness, and lack of sleep; cognitive tasks; emotional state; and physiological phenomena, such as hunger and temperature, tactile stimulation, or voluntary movement. The presence of dystonia can affect motor function, pain/comfort, and ease of care for individuals with CP. The impact of dystonia and its treatment on the respiratory status of patients with CP is unclear as shown by a recent systematic review.[58]

Medication

Although a detailed discussion falls beyond the scope of this review, it is important to note that several types of medications that are commonly used in CP can have an impact on the respiratory system as well (i.e. benzodiazepines, BoNT-A, etc.).[58]

DISCUSSION

From a clinical point of view, patients with CP regularly suffer from respiratory infections. They seem to be prone to a severe or prolonged course of the infection with a higher risk of admission to an intensive care unit.[13] This corresponds to the importance of respiratory diseases in morbidity and mortality in the CP population. However, Proesmans et al.[20] state that the clinical perception of respiratory infections might be overestimated. In their retrospective study, the number of respiratory infections needing treatment or hospital admission was lower than expected. Because their data were gathered retrospectively, this might be an underestimation and their study population consisted of children with profound intellectual disability and multiple disabilities, of which only one in four had a diagnosis of CP.[20]

The latter is a frequently encountered problem. A number of articles describe respiratory problems in patients with 'neurological impairment' and do not study CP as a specific, separate condition, which complicates interpretation of the results. CP, per se, is a heterogeneous disorder,[3] which makes it complex to investigate and may necessitate studying subgroups.

Respiratory morbidity in CP is multifactorial affected. Recurrent aspiration, poor airway clearance, kyphoscoliosis, impaired lung function, and poor nutritional status predispose to respiratory infections. These factors often coexist and interact.[6,11,12,20] When trying to reduce respiratory morbidity, one should address all these aforementioned risk factors as soon as they are established. If, despite adequate preventive strategies, a respiratory infection occurs, it should be treated thoroughly. The combination of both preventive care and adequate treatment of infections may improve the life expectancy of patients with CP.

In caring for patients with CP, preventing respiratory infections is one aspect, but quality of life and comfort are other very important issues. So, when making medical decisions, the ethical aspects should be considered.

Sleep disorders are known to have a negative effect on quality of life and they are more common in patients with CP, so further research on these disorders (particularly obstructive sleep apnea) in patients with CP is worthwhile.[37]

Although the most frequently reported causes of morbidity and mortality in patients with CP are respiratory diseases, the literature concerning this topic is rather scarce and only a limited number of articles have been published in recent years. Problems such as excessive saliva, gastro-oesophageal reflux, and scoliosis are treated as in other patients, but data concerning results in patients with CP and the effect on their respiratory status are scarce or lacking. No data are available on non-acidic reflux, respiratory physiotherapy, prophylactic antibiotic treatment, sputum cultures, and resistance patterns in patients with CP specifically. This indicates the need for further research on these topics in the CP population.

CONCLUSION

Respiratory problems are frequent in the CP population and are an important cause of death. The respiratory status of patients with CP is influenced by recurrent aspiration, impaired airway clearance, deformity of the spine and chest wall, impaired lung function, poor nutritional status, and recurrent respiratory infections. These factors should all be addressed when caring for patients with CP.

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SUPPORTING INFORMATION

The following additional material may be found online:

Table SI: Factors influencing respiratory status in cerebral palsy and treatment options

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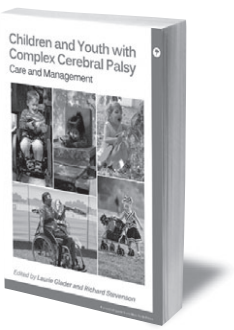

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RESUMEN

MORBILIDAD RESPIRATORIA EN NIÑOS CON PARÁLISIS CEREBRAL: UNA VISIÓN GENERAL

Los problemas respiratorios tienen un impacto significativo sobre la morbilidad y mortalidad en pacientes con parálisis cerebral (PC). En particular, aspiraciones recurrentes, incapacidad para despejar la vía aérea, deformidades de la columna y del tórax, deterioro de la función pulmonar, un pobre estado nutricional e infecciones pulmonares recurrentes, afectan negativamente el estado respiratorio. La displasia broncopulmonar puede contribuir a estos problemas pulmonares, pero el asma no es más común en PC que en la población general. Discutimos las opciones terapéuticas para cada uno de estos factores. La coexistencia e interacción de múltiples factores que influencian el estado respiratorio de las pacientes con PC deben ser reconocidos y tratados efectivamente para reducir la morbilidad respiratoria y la mortalidad.

RESUMO

MORBIDADE RESPIRATÓRIA EM CRIANÇAS COM PARALISIA CEREBRAL: UM PANORAMA

Problemas respiratórios têm impacto significativo na morbidade e mortalidade em pacientes com paralisia cerebral (PC). Em particular, aspiração recorrente, limpeza deficiente das vias aéreas, deformidade espinhal e torácica, função pulmonar deficiente, pobre estado nutricional, e infecções respiratórias recorrentes podem afetar o estado respiratório negativamente. A displasia broncopulmonar pode contribuir para problemas pulmonares, mas a asma não é mais comum em PC do que na população geral. Nós discutimos opções de tratamento para cada um destes fators. Múltiplos fatores coexistentes ou que interagem e que influenciam o estado respiratório de pacientes com PC devem ser reconhecidos e efetivamente abordados para reduzir a morbidade e mortalidade respiratória