Prevention and management of respiratory disease in young people with cerebral palsy: consensus statement

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ABBREVIATIONS

CBR	Consensus-based recommendation
DOSS	Dysphagia Outcome and Severity Scale
EDACS	Eating and Drinking Ability Classification System

Respiratory illness is the leading cause of mortality in children with cerebral palsy (CP). Although risk factors for developing chronic respiratory illness have been identified, comprehensive clinical care recommendations for the prevention and management of respiratory illness do not currently exist. We invited over 200 clinicians and researchers from multiple disciplines with expertise in the management of respiratory illness in children with CP to develop care recommendations using a modified Delphi method on the basis of the RAND Corporation–University of California Los Angeles Appropriateness Method. These recommendations are intended for use by the wide range of practitioners who care for individuals living with CP. They provide a framework for recognizing multifactorial primary and secondary potentially modifiable risk factors and for providing coordinated multidisciplinary care. We describe the methods used to generate the consensus recommendations, and the overall perspective on assessment, prevention, and treatment of respiratory illness in children with CP.

The current life expectancy of young people with very severe cerebral palsy (CP) and CP-like conditions who often have multiple disabilities is much shorter than people without disability. The disabilities in people with CP include severely limited movements over a large part of their bodies, intellectual and communication impairments, and other conditions such as epilepsy, blindness, or deafness.[1]

The major reason for premature death in people with CP relates to respiratory causes.[1,2] The risk of death from respiratory causes for adults with CP is 14 times higher than for adults with no disability.[3] Although vast improvements in medical care and technology occurred in the past 50 years, survival of children with CP has barely altered since the 1970s.[2,4]

In addition to respiratory illnesses being the major contributor of premature death in people with CP, some also have recurrent respiratory illnesses which has a major impact on their quality of life.[5] Children and young people with CP also have a high rate of hospital admissions for respiratory illnesses.[6,7] Hospital care is also very costly. One in four children with CP who go to hospital emergency departments need treatment for respiratory illness.[8] One in eight children with CP who are admitted to hospital need treatment for respiratory illness,[9] and most of them will go on to have another hospital admission for respiratory illness within the same year.[10] Hospital stays for respiratory illness are also 2.5 times as long for children with CP as for other children.[9] Some children stay in hospital for many weeks at a time.[9]

Recent research identifying risk factors for respiratory disease in young people with CP may be used to identify these children earlier.[11–13] However, there is insufficient research evidence, nor any consensus statements among practitioners, on how to prevent or manage respiratory disease in young people with CP.[14] Thus, we developed this consensus statement.

WHAT THIS CONSENSUS STATEMENT CONTAINS

This consensus statement about how respiratory disease in young people with CP should be assessed, prevented, and managed is based on the best available current evidence.

TARGET POPULATION

This consensus statement is for young people with CP and like conditions.[15] The definition of CP is as follows:

> 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.

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> The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior, by epilepsy, and by secondary musculoskeletal problems.[16]

Like conditions are conditions where there is a disturbance of movement and posture because of genetic and metabolic causes. This includes syndromes, brain disorders that do not change, or brain injuries that occur in early childhood.[15]

Not included are any disorders caused by disease or injury of the spine, peripheral nerves, or muscles. Also, any disorders in which nerve or muscle cells deteriorate over time are not included. Nor does this consensus statement include the anaesthetic management of children with CP.

TARGET CONDITIONS

In this consensus statement, respiratory disease in CP may include acute manifestation such as lower respiratory tract infections, with or without hospital admissions, or chronic established disease manifested by the presence of respiratory symptoms such as cough or rattly breathing even when well.

A hospitalization due to respiratory disease is defined as either: (1) a hospitalization to manage the respiratory illness as the primary cause of the hospitalization or (2) a hospitalization episode that is prolonged owing to respiratory illness that develops or is identified during the course of an admission for another reason.

Respiratory disease or hospitalization due to respiratory disease do not include non-recurrent respiratory episodes (e.g. laryngospasm, peri-operative management) or other specific unrelated respiratory conditions (e.g. tracheomalacia).

FRAMEWORK FOR CONSENSUS STATEMENT

This consensus document was written using the framework of the Appraisal of Guidelines for Research and Evaluation II (AGREE II) instrument.[17] The Appraisal consists of six domains covering 23 key items that must be met for the preparation of clinical practice guidelines.

TARGET AUDIENCE FOR THE CONSENSUS STATEMENT

Effective treatment of respiratory illness requires an effective partnership between individuals with CP, their families, carers, and a multidisciplinary team. Thus, we have produced two versions of this consensus statement, covering the following audiences.

The first version is for health professionals (doctors, nurses, dentists, dietitians, physiotherapists, speech pathologists, and occupational therapists). The consensus statement can be used by any clinicians in primary, secondary, tertiary, soun and community health services, teachers, and any individual who works with people with CP. This version aims to inform how to: (1) assess the risk factors for chest infections; (2) manage the risk of chest infections; (3) know when to refer for an assessment of symptoms that are risk factors for chest infections; and (4) make clinical decisions about how to care for children with chest infections.

What this paper adds

- The first consensus statement for preventing and managing respiratory disease in cerebral palsy (CP).
- Risk factors for respiratory disease in CP should be identified early.
- Individuals with CP at risk of respiratory disease require regular assessment of risk factors.
- Effective partnerships between multidisciplinary teams, individuals with CP, and families are essential.
- Treatment of respiratory disease in individuals with CP must be proactive.

The second version is for the CP community, including young people with CP and/or their families and their carers (Appendix S1, online supporting information). This aims to enable people with CP and/or their families to: (1) understand and identify risk factors for chest infections in young people with CP; (2) understand ways of preventing chest infections in young people with CP; (3) know what assessments are recommended; and (4) know what treatments are available for young people with CP with chest infections and why they are recommended.

CONSENSUS STATEMENT COMMITTEE MEMBERS

The full list of the consensus statement committee is given in Appendix S2 (online supporting information). The members adhered to the American Academy of Cerebral Palsy and Developmental Medicine policy for declaration of interest. No committee member had a conflict of interest.

SOURCES OF INFORMATION FOR THE CONSENSUS STATEMENT

This consensus statement is based on three sources of information: (1) systematic review; (2) Delphi study; (3) consumer review.

Systematic review

This was a detailed analysis of all the research papers published from 1998 to 2018 about any treatments aimed at preventing or managing respiratory disease in young people (aged 0–25y) with CP.[14] The systematic review found no high level evidence from controlled trials that any of the interventions were effective in preventing or managing respiratory disease in this population.

Delphi study

This was a standardized and structured technique, using three rounds of questions where clinicians worldwide were asked how they prevent or manage respiratory disease in young people with CP. Details of the Delphi process are outlined in Appendix S3 (online supporting information).

Consumer review

Consumer input was obtained from people living with CP and/or their carers to inform acceptability and utility of

identified prevention and management options. This was a detailed review using interpretive description methodology. A full-length plain language version of the recommendations was submitted to consumers for feedback, and they were subsequently interviewed about the content, format, and dissemination of the document. The consumer version is presented in Appendix S1 with some minor changes since consumer review (including numbering the recommendations and adding a paragraph on advanced care directives). Details of the consumer review process are outlined in Appendix S4 (online supporting information).

GRADING OF THE RECOMMENDATIONS

The recommendations are based on the findings from the systematic review[14] and Delphi study, and the consumer review. They are presented as suggested by the Australian National Health Medical Research Council guide to the development of clinical practice guidelines.[18] The recommendations are classified into three types. The first type is evidence-based recommendations. These are based on the systematic review and show high or moderate support using the Grading of Recommendations, Assessment, Development and Evaluations (GRADE) framework. The second type is the consensus-based recommendations (CBRs). These are recommendations based on agreement among the health professionals who participated in the Delphi study. Third, there are the practice points. These are important practical points that have some published evidence, but participants in the Delphi study did not reach agreement about them.

The consensus statement is presented in three parts: part 1, recognizing and managing risks to prevent respiratory illness in young people with CP; part 2, assessing respiratory health in young people with CP; part 3, treating and managing respiratory illness in young people with CP.

PART 1: RECOGNIZING AND MANAGING RISKS TO PREVENT RESPIRATORY ILLNESS IN YOUNG PEOPLE WITH CP

Recognizing respiratory risk in individuals with CP is everyone's role. Recent research has identified nine risk factors.[13,19,20] A young person with CP who has any of these risk factors is more likely to have at least one hospital admission for chest infections in the next 5 years.

The risk factors are: (1) classification in Gross Motor Function Classification System (GMFCS) level V[21] (in other words, difficulty controlling head and body posture in most positions); (2) at least one hospital admission for respiratory illness in the year preceding the survey; (3) oropharyngeal dysphagia (swallowing difficulties); (4) current seizures; (5) frequent symptoms (having a daily cough, or weekly wheeze, phlegm or gurgly chest); (6) gastro-oesophageal reflux disease; (7) at least two courses of antibiotics for chest infections in the past year; (8) mealtime respiratory symptoms (gurgly voice, wheezing, coughing, sneezing, choking); and (9) snoring every night.

These risk factors are shown in Figure S1 (online supporting information). An interactive online tool has been developed to identify children and young people with CP who have relevant risk factors for developing respiratory disease.[11,13,19,20] The tool can be found at https://www.abilitycentre.com.au/resources/cpchecklist.

There have been no published research trials in the past 20 years showing how to prevent chest infections in typically developing young people with CP.[14] Therefore, the following recommendations are all CBR.

Informing parents early of risk

CBR
1.1. When a child is diagnosed with CP, clinicians should discuss the significance of respiratory illness early and as a priority with the family, particularly when a child is at risk.

Considering risk of respiratory illness based on child's mobility[13]

Different children with CP have different mobility needs. The GMFCS provides an objective classification of gross motor function.[22] The GMFCS can be downloaded free of charge (https://www.canchild.ca/system/tenon/assets/attachments/000/000/058/original/GMFCS-ER_English.pdf).

The GMFCS classifies children into five levels: level I, walks without limitations; level II, walks with limitations in some settings; level III, walks using a hand-held mobility device or uses wheelchair for greater distances; level IV, uses a wheelchair in most settings; level V, uses a wheelchair in all settings and may require additional support for the head or torso.

Children classified in GMFCS level V have a higher risk of chest infections than those in GMFCS levels I to IV.

CBR
1.2. When a child is diagnosed with CP, if they are classified in GMFCS level V, clinicians should discuss with the family the child's increased risk of respiratory illness.

Considering risk of respiratory illness based on swallowing abilities

Children with swallowing difficulties have a greater risk of chest infections than those who can eat and drink without difficulty because they may aspirate food, drink, or saliva, which may lead to respiratory illnesses.

Two assessment tools give information about a person's swallowing abilities: (1) the Dysphagia Outcome and Severity Scale (DOSS);[23] (2) the Eating and Drinking Ability Classification System (EDACS).[24]

The DOSS assesses swallowing difficulties. It classifies children into seven levels, depending on their swallowing abilities: levels 1 and 2 mean a person is unable to take nutrition by mouth; levels 3, 4, and 5 mean a person is able to take nutrition by mouth, but requires thickened

fluids and/or thickened or puréed foods; levels 6 and 7 mean a person is able to eat and drink a normal diet by mouth.

CBR
1.3. When a child is diagnosed with CP, if they are classified in DOSS levels 1, 2, 3, 4, or 5, clinicians should discuss with the family the child's increased risk of respiratory illness.

The EDACS assesses eating and drinking abilities. It has five levels: level I, eats and drinks safely and efficiently; level II, eats and drinks safely but with some limitations to efficiency; level III, eats and drinks with some limitations to safety and possible limitations to efficiency; there maybe limitations to efficiency; level IV, eats and drinks with significant limitations to safety; level V, unable to eat or drink safely—tube feeding may be considered to provide nutrition. The EDACS can be used to assess children only after the age of 3 years. The EDACS can be obtained free of change (https://www.sussexcommunity.nhs.uk/get-involved/research/chailey-research/edacs-request).

CBR
1.4. When a child is diagnosed with CP, if they are classified in EDACS levels III, IV, or V, clinicians should discuss with the family the child's increased risk of respiratory illness.

Identifying and preventing aspiration

Aspiration of foods, drinks, saliva, and refluxate may be noticed when a person coughs or chokes, but most of the time there are no outward signs of aspiration (silent aspiration). Silent aspiration can be detected using a videofluoroscopy.

Aspiration can cause respiratory illness owing to bacteria in the aspirate or chronic inflammatory responses in the lungs.

CBR
1.5. In young people with CP, clinicians should:
1. identify aspiration;
2. assess for comorbidities that increase the risk of aspiration;
3. treat these comorbidities as early as possible.

There are four risk factors for aspiration: (1) dysphagia; (2) uncontrolled seizures; (3) reflux; and (4) drooling saliva (which is a marker of swallowing difficulties). The recommendations for each of these are as follows.

Dysphagia. Clinicians should closely monitor young people with CP who have swallowing difficulties (dysphagia or swallow incoordination) to see whether they are aspirating saliva, food, and/or fluids. Note that aspiration may involve gagging, choking, or coughing, or it may be silent.
CBR
1.6. If young people with CP are suspected of aspirating, clinicians and families should consider a comprehensive assessment by a multidisciplinary team including speech pathology (see part 2).
CBR
1.7. If young people with CP are suspected of aspirating, health professionals and families should consider introducing foods and drinks with thickened textures.
CBR
1.8. If young people with CP are suspected of aspirating, the family and the team of clinicians should consider implementing strategies for developing the person's oral motor skills.
CBR
1.9. If young people with CP have trouble controlling their saliva, clinicians should consider postural support, positioning for head control, and other ways of preventing them from aspirating their saliva.
CBR
1.10. If young people with CP are aspirating, clinicians should develop a schedule for reviewing them regularly.

Uncontrolled seizures. Epilepsy is common in CP. During a seizure, a person may vomit, drool, or stop breathing. The vomit or excess saliva can be aspirated into the lungs. Seizure medications can sometimes cause sedation which leads to increased drooling and further aspiration.
CBR
1.11. Clinicians should monitor young people with CP for ongoing seizures.
CBR
1.12. When seizures become uncontrolled, clinicians should refer young people with CP to a neurologist.

Reflux. Gastro-oesophageal reflux can cause aspiration which in turn may lead to respiratory illnesses.
CBR
1.13. Clinicians should assess and manage gastro-oesophageal reflux disease.

Drooling. Two in five children with CP over the age of 4 years drool with their saliva.[25] This is not because they produce too much saliva, but because they have trouble managing their saliva.[26] Aspiration of saliva leads to ongoing respiratory illnesses.[27,28]
CBR
1.14. Clinicians should assess and manage drooling.
CBR
1.15. If young people with CP are choking on saliva, doctors should review medications that may cause drooling.
CBR
1.16. If young people with CP are drooling with saliva, doctors should consider oral medications, injecting salivary glands with botulinum neurotoxin A, or salivary gland surgery to manage drooling.

There is evidence from uncontrolled trials that salivary gland botulinum neurotoxin A[29–32] and salivary gland surgery may reduce respiratory infections.[33–35]

Optimizing airway clearance

Some children with CP have an ineffective cough and are unable to clear their airways effectively.

CBR
1.17. Physiotherapists should prescribe techniques to maintain clear airways and show carers how to use them.
CBR
1.18. Clinicians should identify ineffective cough and teach families and caregivers techniques to optimize cough to manage secretions. (Refer to 3.9–3.18 for additional recommendations relating to this.)
CBR
1.19. If young people with CP have frequent episodes of a wet cough, clinicians should consider regular chest physiotherapy. (Refer to 3.9–3.18 for additional recommendations relating to this.)

CBR
1.20. If young people with CP have upper airway obstruction, clinicians should manage it, where possible, using positioning and tone management.
CBR
1.21. It is recommended that clinicians and carers optimize positioning for lung expansion.
CBR
1.22. If young people with CP have a history of apnoea, snoring, or if they have large tonsils or swollen turbinates, clinicians should refer them to ear, nose, and throat specialists so that upper airway obstructions can be assessed and managed.
CBR
1.23. If young people with CP are aspirating or at risk of chronic suppurative lung disease, physiotherapists should use preventative measures, such as airway clearance regimes (see 3.19–3.20).

There is evidence from uncontrolled trials that surgery to improve upper airway obstruction improves blood oxygen levels,[36,37] and sleep disturbance and daytime function,[38] but no evidence about its effects on preventing respiratory illness.

CBR
1.24. Clinicians should assist the person with CP and/or instruct their families on techniques to optimize chest wall mobility to prevent restrictive lung disease. This includes the following:
1. physiotherapists prescribing and educating carers in techniques to maintain an individuals' chest mobility;
2. maximizing physical activity and minimizing immobility, including regular position changes for non-ambulant individuals;
3. optimizing management of movement disorders;
4. clinicians assessing individuals for kyphosis, kyphoscoliosis, and lordoscoliosis and initiating a postural plan to prevent or manage these deformities;
5. where surgery for scoliosis is under consideration, a multidisciplinary team evaluating risks and benefits.

Most spinal deformities in children with CP involve neuromuscular scoliosis. However, spinal kyphosis (thoracic spine) and lordosis (lumbar spine) are also common. Spinal deformities in children with CP are related to severity of gross motor function and are most common in GMFCS levels IV and V.[39] Therefore, although scoliosis itself may not be a risk factor for respiratory illness in CP, most of the children with scoliosis fall into the GMFCS functional categories at the highest risk of respiratory illness. There is evidence from one uncontrolled trial that scoliosis surgery did not improve respiratory health.[40]

Optimizing nutrition

There is no research evidence to show whether improving nutritional intake in young people with CP prevents respiratory illness. However, in people without CP, there is evidence that insufficient nutrition leads to weakened immune responses.[41–44]

CBR
1.25. If young people with CP have poor nutritional status, dietitians should optimize nutritional intake.

Optimizing physical activity and fitness

The benefits of exercise for improving respiratory function and fitness are well known and widely accepted. Physical exercise causes deep breathing, and deep breathing helps shift secretions from the airways. Exercise particularly helps to clear smaller airways that cannot be cleared with coughing.[45]

Practice points
1.26. Young people with CP should be as physically active as possible.
Practice points
1.27. Young people with CP should be as fit as possible.

There is evidence from uncontrolled studies that exercise improves pulmonary function in children with CP, but there is no evidence that exercise improves respiratory health.[46–49] There is expert consensus that lung function, physical activity, and fitness should be optimized in young people with CP.

Importance of maintaining dental hygiene

The mouth can be a reservoir for the bacteria that cause respiratory illnesses.[50,51] Among people without CP, those with poor oral health are more likely to develop pneumonia, whereas improving oral health reduces the risk of it.[52] This is likely to be true for people with CP as well.

CBR
1.28. Young people with CP should receive regular dental care.

Regular dental care includes education to carers about good dental hygiene including daily dental care and review with a dentist (twice a year).[53]

Vaccination against influenza

CBR
1.29. Young people with CP and their families should be vaccinated annually against influenza.

Reducing exposure to tobacco smoke

CBR
1.30. Young people with CP and their families should avoid exposure to tobacco smoke.

Managing asthma

CBR
1.31. Clinicians should be alert for asthma in young people with CP.
CBR
1.32. Doctors should assess the responses of young people with CP to asthma medications. If symptoms do not improve, then doctors should consider stopping the medication.

Ongoing surveillance for risk of respiratory disease

The major risk factors for respiratory disease in young people with CP are known and most of them can be treated. Regular screening for these risk factors could, therefore, prevent many respiratory illnesses in these young people.

CBR
1.33. Young people with CP should be screened for risk of respiratory disease at least every 12 months if they meet any of the following criteria:
1. a hospital admission for respiratory illness in the past 12 months;
2. GMFCS level V;
3. EDACS level III–V (for children over 3y) or based on DOSS (for children under 3y).

There was no consensus reached about the frequency of respiratory surveillance. However, recommendations for increased respiratory review under specific situations below were reached.

CBR
1.34. Clinicians should consider screening young people with CP for risk of respiratory disease more often if they meet any of the following criteria:
1. an increase in hospital admissions for respiratory illnesses;
2. a hospital admission for a respiratory illness since last review;
3. poor control of seizures;
4. significant change in nutritional status;
5. a change from oral intake to tube feeding;
6. evidence of aspiration (from clinical swallow assessment, increased choking episodes, chest X-ray, and/or videofluoroscopy);
7. deterioration in gross motor function, particularly decreased ability, or tolerance of sitting or standing;
8. deterioration in oromotor function;
9. difficulty managing secretions;
10. presence of kyphosis, kyphoscoliosis, or lumboscoliosis;
11. other changes in clinical status affecting ability to manage and clear secretions, e.g. illness, pain, fatigue;
12. any other respiratory-related concern identified by the family or clinician.

PART 2: ASSESSMENT OF RESPIRATORY HEALTH IN CHILDREN WITH CP

Respiratory review is an ongoing process that continues for every child until the risk for respiratory illness is mitigated. Respiratory review will assist in identification of both risk and presence of respiratory illness, inform planning for ongoing management of respiratory risk factors, support education, and assist clear communication.

Initial assessments

Clinicians should assess young people with CP for their risk of respiratory disease. The assessment includes a thorough history and clinical assessment (when possible). It aims to identify and document concerns, risk factors, care and comfort needs, and changes in gross motor function or eating and swallowing function. The assessor should be able to identify the child's GMFCS and EDACS levels and, where possible, refer to a speech pathologist for evaluation of DOSS.

CBR
2.1. Clinical assessments should be multidisciplinary.
CBR
2.2. Clinicians should systematically assess symptoms of respiratory disease.
CBR
2.3. Clinicians should obtain a detailed history that includes all aspects of risk or comorbidities as far as they relate to the risk of respiratory disease:
1. GMFCS;
2. EDACS or DOSS by a speech pathologist if under 3 years of age;
3. epilepsy;
4. gastro-oesophageal reflux disease;
5. drooling;
6. episodes of aspiration, wheezing, or noisy breathing.

CBR
2.4. Clinicians should obtain a history of previous respiratory illnesses during the last 12 months with reference to:
1. antibiotic use (frequency, type, and duration);
2. hospital admissions for respiratory illnesses;
3. hospital admissions where the young person with CP developed a respiratory illness after being admitted to hospital; and
4. need and use of non-invasive ventilation during the past 12 months.

CBR
2.5. The clinical assessment should include physical examination of breathing pattern (rhythm, depth, pattern of chest wall movement), respiratory rate, heart rate, work of breathing, colour, chest wall shape, palpation, auscultation, visualization of tonsils, and oxygen saturations. (This needs to be done with the young person with CP when in good health. When the young person becomes sick with a respiratory illness, it will be used as a baseline for comparison.)
CBR
2.6. Clinicians should assess how well the young person with CP is able to manage secretions. This should be done when the young person is well and again when the young person is unwell.
CBR
2.7. Clinicians should assess nutritional status (weight, height, rate of growth, blood test, dietitian review).
CBR
2.8. Clinicians should consider the medical stability of a young person with CP before initiating feed trials.
CBR
2.9. Assessment of eating and drinking abilities and risk of aspiration in a young person with CP should be multidisciplinary (e.g. speech pathologists, occupational therapists, physiotherapists, and dietitians). This should be done when the young person is well and again when the young person is unwell.
1. Where swallowing difficulties are suspected, speech pathologists should assess oropharyngeal motor dysfunction.
2. Swallowing using a range of different food textures and different drink textures should be assessed.

CBR
2.10. Clinicians should assess young people with CP for skeletal deformities, including kyphosis, kyphoscoliosis, and lumboscoliosis.

Referrals for diagnostic tests

When the assessments above have been completed, clinicians may refer a young person with CP for further tests, as follows.

CBR
2.11. A videofluoroscopy should be performed on young people with CP when clinical history/clinical observations/bedside assessment indicates that there may be a risk of aspiration.

CBR
2.12. Clinicians should consider a sleep study (including assessment of overnight oxygen saturation) for young people with CP with symptoms of obstruction or apnoea.
Practice points
2.13. Sputum culture, chest imaging, and computed tomography of the chest may provide additional information to guide treatments in some instances.

PART 3: TREATMENT OF RESPIRATORY ILLNESS IN CP

Treatment of respiratory illness, like prevention, involves a partnership between health professionals, young people with CP, and families of young people with CP. Management must be proactive and timely. The following recommendations are for treatment of young people with CP with respiratory illnesses.

General recommendations for management of respiratory illness

When a young person with CP has a respiratory illness, we recommend the following.

CBR
3.1. All the recommendations from part 1 apply to treatment of respiratory illness too.
CBR
3.2. Clinicians should consider early active investigation and a low threshold for treatment of any sign of respiratory disease.
Practice points
3.3. The roles and responsibilities of the multidisciplinary team need to be determined, including allocating a team leader who takes responsibility to make decisions that inform the direction of treatment, and coordinate care decisions with the young person with CP and their family.
CBR
3.4. Clinicians should guide interventions on the basis of whether the respiratory illness is a community-acquired infection, a hospital-acquired infection, or whether aspiration is the cause.

Use of medications

Medications to manage respiratory illnesses considered in the Delphi study included antibiotics, bronchodilators, anti-inflammatory agents to manage airway inflammation and bronchospasm, and mucolytics to help thin secretions. Only the following medicinal recommendations for treating young people with CP with respiratory illnesses reached consensus.

CBR
3.5. As soon as a young person with CP gets a lower respiratory tract infection, clinicians should treat it with antibiotics using antibiotic guidelines.
CBR
3.6. When young people with CP have respiratory illnesses, clinicians ensure that excessive drooling of saliva is managed, but consider that interventions that thicken airway secretions may cause mucus plugging.

There was no evidence identified in the systematic review and no consensus about the efficacy of mucolytic agents for treating respiratory illness in young people with CP.

There is evidence from published case studies of two children with CP that nebulized tobramycin may reduce the number of episodes of pneumonia, number of hospital admissions, and length of stay.[54]

Optimizing general health

CBR
3.7. When young people with CP have respiratory illnesses, clinicians should consider whether nutritional status is sufficient to aid recovery from infection.
CBR
3.8. When young people with CP have respiratory illnesses, clinicians should ensure any gastro-oesophageal reflux disease is managed.

There are no studies to show whether positioning or medications to manage reflux improves respiratory disease in young people with CP.

Optimizing airways

If upper airway obstruction is multilevel, or if there are day and night symptoms, this may require specialist, multidisciplinary input in consultation with the young person with CP and/or their family. It is important to not miss easily remediable causes such as adeno-tonsillar hypertrophy or iatrogenic hypotonia.

There was no consensus reached on the use of non-invasive ventilation or surgery to manage upper airway obstruction. There is published evidence that surgery for upper airway obstruction in young people with CP improves blood oxygen levels,[36,37] and improves sleep and the ability to function during the daytime.[38] However, none of these studies compared the children who had surgery with those without surgery.

Physiotherapy for airway clearance therapy

Owing to the limited number of published studies for airway clearance therapy in young people with CP, the need to carefully monitor each individual's response to treatments, both positive and negative, is essential.

CBR
3.9. When young people with CP have symptoms of respiratory disease or sputum retention, clinicians should consider referring them to a physiotherapist with experience treating respiratory problems in children with CP.
CBR
3.10. When young people with CP have respiratory illnesses, physiotherapists should prescribe changes to routine positioning to optimize lung function, and educate carers on how to do this.
CBR
3.11. When young people with CP have respiratory illnesses, physiotherapists should assess the strength and effectiveness of the cough for clearing secretions.
CBR
3.12. When young people with CP have respiratory illnesses, physiotherapists should show carers how to position them to improve effectiveness of coughing. This includes providing good support for the neck and spine.

CBR
3.13. When young people with CP have respiratory illnesses, if they have an ineffective cough but adequate control of the upper airway, physiotherapists should show carers how to assist their cough manually.
CBR
3.14. When young people with CP have respiratory illnesses, if non-invasive methods for clearing secretions are ineffective, and/or they cannot swallow safely, clinicians should show families how to suction.
CBR
3.15. When young people with CP have respiratory illnesses, if they have a productive cough, chest physiotherapy should be prescribed, ensuring the person with CP can manage evacuation of secretions safely.
CBR
3.16. When young people with CP have respiratory illnesses, if they have a chronic wet/productive cough, physiotherapists should prescribe long-term daily airway clearance regimes.
CBR
3.17. When young people with CP have respiratory illnesses and are on airway clearance regimens, physiotherapists should monitor and change these regimens according to changing individual respiratory needs.
CBR
3.18. When young people with CP have respiratory illnesses and are on airway clearance regimens, physiotherapists should be alert to, and monitor adverse events of, these regimens.

Although there was consensus about positioning to optimize lung function, there is no research evidence for the effectiveness of this intervention for respiratory health.[55,56]

There was no consensus reached on the use of mechanical assistive devices, such as mechanical insufflation-exsufflation or high-frequency chest-wall oscillation to augment secretion clearance. There is evidence from uncontrolled trials that high-frequency chest-wall oscillation reduces respiratory hospital days, episodes of pneumonia, and days on antibiotics,[57–60] but a randomized controlled study failed to find any evidence of an effect on hospital days or antibiotics because of its small sample size (only nine of the participants were young people with CP).[61] There is no current evidence that mechanical insufflation–exsufflation makes a difference to hospital length of stay, days on oxygen, or days of physiotherapy in young people with CP.[62]

Reviewing mealtime management plans

CBR
3.19. When young people with CP have respiratory illnesses or any other illness, speech pathologists should reassess dysphagia as the risk of aspiration may increase when the child is unwell.
CBR
3.20. When young people with CP have respiratory illnesses, and a swallowing assessment shows that alternatives to oral intake are indicated, this is discussed with the young person with CP, family, medical team, and dietitian.

There is evidence from an uncontrolled trial that providing mealtime management advice to mothers of children with CP reduces chest infections.[63,64] There is also evidence from an uncontrolled trial that multidisciplinary management guided by assessment using videofluoroscopy reduces the number of exacerbations and the number of visits to the emergency department.[65]

There is evidence from an uncontrolled trial that thickening enteral feeds reduces cough, wheeze, and reflux.[66] There is evidence from uncontrolled trials that respiratory illnesses are improved by gastrostomy with or without fundoplication.[67–70] One uncontrolled trial reported no difference.[71] Several of these studies also reported significant adverse events after these treatments, although not necessarily caused by the treatments. It should be noted that the children receiving these treatments usually have very complex conditions and are at high risk of adverse outcomes when unwell.

Advanced care planning

Clinical deterioration towards death is not always a steady decline and the individual's health can fluctuate.[72] We are not good at predicting timing of death, although evidence shows that there is an increase in hospital admissions in the 6 months before death.[73] The benefits of referring to palliative care team are well researched.[74–76] Consideration of major surgical interventions such as a tracheostomy to support breathing or repeated admissions to intensive care units owing to respiratory failure often leads clinicians and families to refocus goals of care to comfort measures. Some families want to embark upon every available treatment option. Clear and honest discussion with palliative care or an ethical consultation can be helpful. Depending on preferences, some clinicians and families refocus goals of care before this point. Documentation of these discussions, even when no agreements are reached, is important.

STRENGTHS AND LIMITATIONS OF THE CONSENSUS STATEMENT

This consensus statement was based on limited published evidence supplemented by agreement among health professionals and families of young people with CP. Its strengths are that it is targeted specifically at young people with CP. Also, it is informed by professionals from many disciplines, including those who provide care for young people with CP and researchers who investigate the best ways to provide this care. Third, it is guided by consumer input. Finally, it takes into account specific individual risk factors. Its major limitation is that it lacks a strong evidence base.

Each young person with CP will respond differently to the treatments described in this consensus statement. Each young person's response to the treatments should be assessed carefully. In clinical practice, treatments should be flexible and take into account individual needs and circumstances, and monitor very carefully for adverse reactions.

We appreciate that clinicians want much more detail than it has been possible to provide in this consensus statement. The statement is not our own; it represents what was agreed upon by clinicians and researchers working in this field.

This is the very first set of guidelines issued for the prevention and/or management of respiratory disease in young people with CP. In its current state, its use may be largely in revealing how much we do not know about how to

prevent and manage the leading cause of death in this population, and therefore in stimulating and focusing researchers and clinicians to gather evidence and start filling in the many gaps identified here.

FUTURE RESEARCH

The systematic review showed that there was little good quality research to inform recommendations and form a basis for health-care policy. Many studies did not have comparison groups and good outcome measures. Few studies investigated patients' experiences of treatments, quality of life, or cost effectiveness.

There is wide scope for future research into preventing and managing respiratory disease in young people with CP. The systematic review and Delphi study both uncovered a wide diversity of treatments believed to benefit young people with CP with chest infections but very little evidence for any of these in CP (although many of these treatments have been used successfully with other populations).

Treatments that need further research are influenza and other respiratory vaccinations; prophylactic or early use of antibiotics; conventional physiotherapy techniques; airway clearance techniques; exercise; positioning; non-invasive ventilation—tolerance and efficacy in CP; dental health; pharmaceutical and surgical methods of reducing drooling with saliva; surgical management of upper airways; mealtime management; oropharyngeal muscle training; electrical stimulation of oropharyngeal muscles; nutritional management; management of reflux; gastro-intestinal surgery; spinal surgery, and management of seizures.

Concurrently with research on these interventions, there needs to be research on the causes and progression of respiratory disease over time.

All current published evidence examined single treatments rather than multiple treatments for respiratory disease in young people with CP. Multiple treatments are used in clinical practice. Therefore, it is recommended that future research examine combinations of treatments and care models.

There are no studies on interventions for the prevention of respiratory disease in young people with CP who are at risk. Now that risk factors are known,[12,13,19] it is recommended that preventative interventions for these children be explored.

All future research on chest infections in CP needs to be well designed, high quality, with sufficient sample sizes and follow-up periods to determine whether treatments are effective. Future research also needs to include patient-reported outcomes (e.g. activities, participation, and quality of life).

DISCLAIMER AND REVIEW OF THE RECOMMENDATIONS

This consensus is based on a systematic review of the current literature published in 2019[14] and careful and considered analysis of expert opinion achieved by a Delphi process conducted in 2019. It is provided as guidance.

There may well be a range of unknown factors yet to be determined in the assessment and management of respiratory illness in people with CP. Those factors, and the assessment of the individual patient, may affect the assessment of the appropriate clinical treatment for an individual patient. Clinical judgement can and should override these recommendations when clinical or carer concerns are noted, and appropriate action taken to meet the needs of the individual patient.

The consensus statement is to be reviewed within 5 years from the date of publication to assess uptake and impact of the recommendations, and to review new knowledge that may affect the recommendations.

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SUPPORTING INFORMATION

The following additional material may be found online:

Appendix S1: Plain language version.

Appendix S2: Consensus panel for the prevention and management of respiratory disease in young people with cerebral palsy.

Appendix S3: The Delphi process to achieve consensus for the prevention and management of respiratory disease in young people with cerebral palsy: standards of care.

Appendix S4: Consumer review process of the consensus statement for the prevention and management of respiratory disease in young people with cerebral palsy.

Figure S1: Risk factors for respiratory hospital admissions for young people (1–26y) with cerebral palsy.

REFERENCES

1. Blair E, Langdon K, McIntyre S, Lawrence D, Watson L. Survival and mortality in cerebral palsy: observations to the sixth decade from a data linkage study of a total population register and National Death Index. *BMC Neurol* 2019; **19**: 111.
2. Reid SM, Carlin JB, Reddihough DS. Survival of individuals with cerebral palsy born in Victoria, Australia, between 1970 and 2004. *Dev Med Child Neurol* 2012; **54**: 353–60.
3. Ryan JM, Peterson MD, Ryan N, et al. Mortality due to cardiovascular disease, respiratory disease, and cancer in adults with cerebral palsy. *Dev Med Child Neurol* 2019; **61**: 924–8.
4. Himmelmann K, Sundh W. Survival with cerebral palsy over five decades in Western Sweden. *Dev Med Child Neurol* 2015; **57**: 762–7.
5. Elema A, Zalmstra TA, Boonstra AM, Narayanan UG, Reinders-Messelink HA, Putten VD. Pain and hospital admissions are important factors associated with quality of life in nonambulatory children. *Acta Paediatr* 2016; **105**: e419–e25.
6. Murphy NA, Hoff C, Jorgensen T, Norlin C, Young PC. Costs and complications of hospitalizations for children with cerebral palsy. *Pediatr Rehabil* 2006; **9**: 47–52.
7. Young NL, McCormick AM, Gilbert T, et al. Reasons for hospital admissions among youth and young adults with cerebral palsy. *Arch Dis Child* 2011; **92**: 46–50.

8. Meehan E, Reid SM, Williams K, et al. Tertiary paediatric emergency department use in children and young people with cerebral palsy. *J Paediatr Child Health* 2015; **51**: 994–1000.
9. Meehan E, Reid SM, Williams K, et al. Hospital admissions in children with cerebral palsy: a data linkage study. *Dev Med Child Neurol* 2017; **59**: 512–9.
10. Meehan E, Freed GL, Reid SM, et al. Tertiary paediatric hospital admissions in children and young people with cerebral palsy. *Child Care Health Dev* 2015; **1**: 928–37.
11. Blackmore AM, Bear N, Blair E, et al. Prevalence of symptoms associated with respiratory illness in children and young people with cerebral palsy. *Dev Med Child Neurol* 2016; **58**: 780–1.
12. Blackmore AM, Bear N, Blair E, et al. Predicting respiratory hospital admissions in young people with cerebral palsy. *Arch Dis Child* 2018; **103**: 1119–24.
13. Blackmore AM, Bear N, Langdon K, Moshovis L, Gibson N, Wilson A. Respiratory hospital admissions and emergency department visits in young people with cerebral palsy: 5-year follow-up. *Arch Dis Child* 2019; 1–2. https://doi.org/10.1136/archdischild-2019-317714
14. Blackmore AM, Gibson N, Cooper MS, Langdon K, Moshovis L, Wilson AC. Interventions for management of respiratory disease in young people with cerebral palsy: A systematic review. *Child Care Health Dev* 2019; **45**: 754–71.
15. Smithers-Sheedy H, Badawi N, Blair E, et al. What constitutes cerebral palsy in the twenty-first century? *Dev Med Child Neurol* 2014; **56**: 323–8.
16. Rosenbaum P, Paneth N, Leviton A, et al. A report: the definition and classification of cerebral palsy April 2006. *Dev Med Child Neurol* 2007; **109**: 8–14.
17. Brouwers M, Kho M, Browman G, et al. AGREE II: advancing guideline development, reporting and evaluation in healthcare. *Can Med Assoc J* 2010; **182**: E839–42.
18. National Health Medical Research Council. Guide to the development, implementation and evaluation of clinical practice guidelines. Canberra, Australia: National Health Medical Research Council, 2009.
19. Blackmore AM, Bear N, Blair E, et al. Factors associated with respiratory illness in children and young adults with cerebral palsy. *J Pediatr* 2016; **168**: 151–7.
20. Bear N, Blackmore A, Gibson N, et al. Validation of factors associated with respiratory hospitalizations in young people with cerebral palsy. *Dev Med Child Neurol* 2018; **60**(Suppl 1): 50.
21. Palisano R, Rosenbaum P, Bartlett D, Livingston M. Gross Motor Function Classification System–expanded and revised [Internet]. Available at https://www.canchild.ca/system/tenon/assets/attachments/000/000/058/original/GMFCS-ER_English.pdf (accessed 20 January 2020).
22. Palisano R, Hanna S, Rosenbaum P, Russell D. Validation of a model of gross motor function for children with cerebral palsy. *Phys Ther* 2000; **80**: 974–85.
23. O'Neil KH, Purdy M, Falk J, Gallo L. The dysphagia outcome and severity scale. *Dysphagia* 1999; **14**: 139–45.
24. Sellers D, Mandy A, Pennington L, Hankins M, Morris C. Development and reliability of a system to classify the eating and drinking ability of people with cerebral palsy. *Dev Med Child Neurol* 2014; **56**: 245–51.
25. Reid SM, McCutcheon J, Reddihough DS, Johnson H. Prevalence and predictors of drooling in 7- to 14-year-old children with cerebral palsy: a population study. *Dev Med Child Neurol* 2012; **54**: 1032–6.
26. Erasmus CE, Van Hulst K, Rotteveel LJC, et al. Drooling in cerebral palsy: hypersalivation or dysfunctional oral motor control? *Dev Med Child Neurol* 2009; **51**: 454–9.
27. Erasmus CE, Van Hulst K, Rotteveel JJ, Willemsen MAAP, Jongerius PH. Swallowing problems in cerebral palsy. *Eur J Pediatr* 2012; **171**: 409–14.
28. Owayed AF, Campbell DM, Wang EEL. Underlying causes of recurrent pneumonia in children. *Arch Pediatr Adolesc Med* 2000; **154**: 190–4.
29. Faria J, Harb J, Hilton A, Yacobucci D, Pizzuto M. Salivary botulinum toxin injection may reduce aspiration pneumonia in neurologically impaired children. *Int J Pediatr Otorhinolaryngol* 2015; **79**: 2124–8.
30. Gubbay A, Blackmore AM. Effects of salivary gland botulinum toxin-A on drooling and respiratory morbidity in children with neurological dysfunction. *Int J Pediatr Otorhinolaryngol* 2019; **124**: 124–8.
31. Kim H, Lee Y, Weiner D, Kaye R, Cahill AM, Yudkoff M. Botulinum toxin type A injections to salivary glands: combination with single event multilevel chemoneurolysis in 2 children with severe spastic quadriplegic cerebral palsy. *Arch Phys Med Rehabil* 2006; **87**: 141–4.
32. Meece RW, Fishlock KF, Bayley EW, Keller MS. Ultrasound-guided botox injections of salivary glands in children with drooling. *J Radiol Nurs* 2010; **29**: 20–4.
33. Manrique D, Sato J. Salivary gland surgery for control of chronic pulmonary aspiration in children with cerebral palsy. *Int J Pediatr Otorhinolaryngol* 2009; **73**: 1192–4.
34. Noonan K, Prunty S, Ha JF, Vijayasekaran S. Surgical management of chronic salivary aspiration. *Int J Pediatr Otorhinolaryngol* 2014; **78**: 2079–82.
35. Vijayasekaran S, Unal F, Schraff SA, Johnson RF, Rutter MJ. Salivary gland surgery for chronic pulmonary aspiration in children. *Int J Pediatr Otorhinolaryngol* 2007; **71**: 119–23.
36. Hartzell LD, Guillory RM, Munson PD, Dunham AK, Bower CM, Richter GT. Tongue base suspension in children with cerebral palsy and obstructive sleep apnea. *Int J Pediatr Otorhinolaryngol* 2013; **77**: 534–7.
37. Myatt HM, Beckenham EJ. The use of diagnostic sleep nasendoscopy in the management of children with complex upper airway obstruction. *Clin Otolaryngol Allied Sci* 2000; **25**: 200–8.
38. Hsiao KH, Nixon GM. The effect of treatment of obstructive sleep apnea on quality of life in children with cerebral palsy. *Res Dev Disabil* 2007; **29**: 133–40.
39. McMillan LE, Dudhniwala AG, Ahuja S. Is there a correlation between gross motor function classification system (GMFCS) level and scoliosis in patients with cerebral palsy (CP)? A systematic review. *Spine J* 2016; **16**: S55.
40. Keskinen H, Lukkarinen H, Korhonen K, Jalanko T, Koivusalo A, Helenius I. The lifetime risk of pneumonia in patients with neuromuscular scoliosis at a mean age of 21 years: the role of spinal deformity surgery. *J Child Orthop* 2015; **9**: 357–64.
41. Blumentals WA, Nevitt A, Peng MM, Toovey S. Body mass index and the incidence of influenza-associated pneumonia in a UK primary care cohort. *Influenza Other Respir Viruses* 2011; **6**: 28–36.
42. Ritz BW, Gardner EM. Malnutrition and energy restriction differentially affect viral immunity. *J Nutrit* 2006; **136**: 1141–4.
43. Scrimshaw NS, SanGiovanni JP. Synergism of nutrition, infection, and immunity: an overview. *Am J Clin Nutr* 1997; **66**: 464S–77S.
44. dos Santos Souza, Simon MI, Drehmer M, de Abreu e Silva FA et al. Association of nutritional status, plasma, albumin levels and pulmonary function in cystic fibrosis. *Nutricion Hospitalaria* 2011; **26**: 1322–7.
45. Braverman J. Respiratory problems in individuals with cerebral palsy: recognition, management and prevention. *Exceptional Parent* 2001; **31**: 56–8.
46. Hutzler Y, Chacham A, Bergman U, Szeinberg A. Effects of a movement and swimming program on vital capacity and water orientation skills of children with cerebral palsy. *Dev Med Child Neurol* 1998; **40**: 176–81.
47. Rothman JG. Effects of respiratory exercises on the vital capacity and forced expiratory volume in children with cerebral palsy. *Physical Therapy* 1978; **58**: 421–5.
48. Shin SO, Kim NS. Upper extremity resistance exercise with elastic bands for respiratory function in children with cerebral palsy. *J Phys Ther Sci* 2017; **29**: 2077–80.
49. Lee HY, Cha YJ, Kim K. The effect of feedback respiratory training on pulmonary function of children with cerebral palsy: a randomized controlled preliminary report. *Clin Rehabil* 2014; **28**: 965–71.
50. Scannapieco FA, Bush RB, Paju S. Associations between periodontal disease and risk for nosocomial bacterial pneumonia and chronic obstructive pulmonary disease: a systematic review. *Ann Periodontol* 2003; **8**: 54–69.
51. Mojon P. Oral health and respiratory infection. *J Can Dent Assoc* 2002; **68**: 340–5.
52. Azarpazhooh A, Leake JL. Systematic review of the association between respiratory diseases and oral health. *J Periodontol* 2006; **77**: 1465–82.
53. Norwood K, Slayton R. Oral health care for children with developmental disabilities. *Pediatrics* 2013; **131**: 614–9.
54. Plioplys AV, Kasnicka I. Nebulized tobramycin: prevention of pneumonias in patients with severe cerebral palsy. *J Pediatr Rehabil Med Interdiscip Appr* 2011; **4**: 155–8.
55. Lephart K, Kaplan SL. Two seating systems' effects on an adolescent with cerebral palsy and severe scoliosis. *Pediatr Phys Ther* 2015; **27**: 258–66.
56. Littleton SR, Heriza CB, Mullens PA, Moerchen VA, Bjornson K. Effects of positioning on respiratory measures in individuals with cerebral palsy and severe scoliosis. *Pediatr Phys Ther* 2011; **23**: 159–69.
57. Fitzgerald K, Dugre J, Pagala S, Homel P, Marcus M, Kazachkov M. High-frequency chest wall compression therapy in neurologically impaired children. *Resp Care* 2014; **59**: 107–12.
58. Garuti G, Verucchi E, Fanelli I, Giovannini M, Winck JC, Lusuardi M. Management of bronchial secretions with free aspire in children with cerebral palsy: impact on clinical outcomes and healthcare resources. *Ital J Pediatr* 2016; **42**: 7.

59. Plioplys AV. Pulmonary vest therapy in pediatric long-term care. *J Am Med Dir Assoc* 2002; **3**: 318–21.
60. Plioplys AV, Ebel J, Kasnicka I. Pulmonary vest therapy to prevent pneumonia in quadriplegic cerebral palsy. *Arch Phys Med Rehabil* 2003; **84**: E6.
61. Yuan N, Kane P, Shelton K, Matel J, Becker BC, Moss RB. Safety, tolerability, and efficacy of high-frequency chest wall oscillation in pediatric patients with cerebral palsy and neuromuscular diseases: an exploratory randomized controlled trial. *J Child Neurol* 2010; **25**: 815–21.
62. 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; **63**: 187–93.
63. Adams MS. The management of feeding difficulties in children with cerebral palsy in Bangladesh. London, UK: University College London, 2009.
64. Adams MS, Khan NZ, Begum SA, Wirz SL, Hesketh T, Pring TR. Feeding difficulties in children with cerebral palsy: low-cost caregiver training in Dhaka, Bangladesh. *Child Care Health Dev* 2012; **38**: 878–88.
65. Romero RG, Arnal IG, Montañés MJR, et al. Evaluación de la disfagia. Resultados tras un año de la incorporación de la videofluoroscopia en nuestro centro. *An Pediatr (Barc)* 2017; **89**: 92–7.
66. Miyazawa R, Tomomasa T, Kaneko H, Arakawa H, Shimizu N, Morikawa A. Effects of pectin liquid on gastroesophageal reflux disease in children with cerebral palsy. *BMC Gastroenterol* 2008; **8**: 11.
67. Ishimaru Y. Efficacy of laparoscopic fundoplication for gastroesophageal reflux disease in neurologically impaired patients: postoperative quality of life and operative outcomes. *Dokkyo J Med Sci* 2017; **44**: 141–50.
68. O'Loughlin EV, Somerville H, Shun A, et al. Antireflux surgery in children with neurological impairment: caregiver perceptions and complications. *J Pediatr Gastroenterol Nutr* 2013; **56**: 46–50.
69. Sullivan PB, Juszczak E, Bachlet AM, et al. Gastrostomy tube feeding in children with cerebral palsy: a prospective, longitudinal study. *Dev Med Child Neurol* 2005; **47**: 77–85.
70. Sullivan PB, Morrice JS, Vernon-Roberts A, Grant H, Eltumi M, Thomas AG. Does gastrostomy tube feeding in children with cerebral palsy increase the risk of respiratory morbidity? *Arch Dis Child* 2006; **91**: 478–82.
71. Cheung KM, Tse HW, Tse PWT, Chan KH. Nissen fundoplication and gastrostomy in severely neurologically impaired children with gastroesophageal reflux. *Hong Kong Med J* 2006; **12**: 282–8.
72. Feudtner C. Collaborative communication in pediatric palliative care: a foundation for problem-solving and decision-making. *Pediatr Clin North Am* 2007; **54**: 583–607.
73. Vemuri S, Baker L, Williams K, Hynson J. The last 2 years of life for children with severe physical disability: Observations from a tertiary paediatric centre. *J Paediatr Child Health* 2018; **54**: 1357–61.
74. Vickers J, Thompson A, Collins GS, Childs M, Hain R, Paediatric Oncology Nurses' Forum/United Kingdom Children's Cancer Study Group Palliative Care Working Group. Place and provision of palliative care for children with progressive cancer: a study by the Paediatric Oncology Nurses' Forum/United Kingdom Children's Cancer Study Group Palliative Care Working Group. *J Clin Oncol* 2007; **25**: 4472–6.
75. Abernethy AP, Currow DC, Fazekas BS, Luszcz MA, Wheeler JL, Kuchibhatla M. Specialized palliative care services are associated with improved short- and long-term caregiver outcomes. *Support Care Cancer* 2008; **16**: 585–97.
76. Hammes BJ, Klevan J, Kempf M, Williams MS. Pediatric advance care planning. *J Palliat Med* 2005; **8**: 766–73.