

CONSENSUS STATEMENT

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

CONSUMER VERSION: FOR CONSUMERS AND FAMILIES



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REASON FOR THIS CONSENSUS STATEMENT

Most young people with cerebral palsy (CP) can expect to live about as long as people with no disability. However, for young people with very severe CP and multiple disabilities life expectancy is very much shorter. This includes young people with severely limited movements over a large part of their bodies, limited understanding and communication, and other conditions such as epilepsy, blindness, or deafness.¹

When people with CP die younger than expected, it is usually because of respiratory disease.^{1,2} The risk of death from respiratory disease for adults with CP is 14 times higher than for adults with no disability.³ Children and young people with CP also have a high rate of hospital admissions for respiratory illnesses.^{4,5} The past 50 years have seen vast improvements in medical care and technology. But survival of children with CP is much the same as it was in the 1970s.^{6,7}

Some young people with CP have repeated respiratory illnesses, and this has a major impact on their quality of life.⁸ Hospital care is also very costly. One in four children with CP who go to hospital emergency departments need treatment for respiratory illnesses.^{9,10} One in eight children with CP who are admitted to hospital need treatment for respiratory illnesses,¹¹ and most of them will go on to have another hospital admission for respiratory illnesses within the same year.¹² Hospital stays for respiratory illnesses are also 2.5 times **as long for children with CP as for other children**.¹¹ Some children stay in hospital for many weeks at a time.¹¹

Recently researchers have worked out the risk factors for respiratory disease in young people with CP. These risk factors can be used to identify these children earlier.¹³⁻¹⁵ But there are no guidelines on how to prevent or manage respiratory disease in young people with CP.

WHAT THIS CONSENSUS STATEMENT CONTAINS

This Consensus Statement contains the best available evidence about how respiratory disease in young people with CP should be assessed, prevented, and managed.

TARGET POPULATION

This Consensus Statement is for young people with **CP and like conditions**.¹⁶ 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. 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.” (p. 9, Rosenbaum et al. 2007).¹⁷

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 don't change, or brain injuries that occur in early childhood.¹⁶

Not included are any disorders caused by disease or injury of the spine, peripheral nerves, or muscles. Also not included are any disorders in which nerve or muscle cells deteriorate over time. This Consensus Statement also does not include anaesthetic management of children with CP.

TARGET CONDITIONS

In this Consensus Statement, **respiratory disease** includes acute cough (or increase or change in character of chronic cough), fever, rapid breathing, difficulty breathing, or shortness of breath. Children may appear unwell or sluggish. Some children may have less oxygen in their blood than usual or they may need additional support to help them breathe. Respiratory illnesses may occur once, or they may occur again and again.

A **hospital admission for a respiratory illness** can happen either when a child is admitted with a respiratory illness or when a child is admitted for another reason, but needs to stay in hospital longer because he or she develops a respiratory illness.

Respiratory disease (in this Consensus Statement) does not include once-off breathing problems with a cause above the level of the chest (e.g. tonsillitis, spasm of the vocal cords, management of breathing around surgery) or other specific conditions (e.g., floppy windpipe).

FRAMEWORK FOR THIS CONSENSUS STATEMENT

This consensus document was written using the framework of the Appraisal of Guidelines for Research and Evaluation II (AGREE II) instrument.¹⁸ The AGREE II consists of six domains covering 23 key items that must be met for the preparation of clinical practice guidelines.

TARGET AUDIENCE FOR THIS CONSENSUS STATEMENT

There are two versions of this Consensus Statement:

1. a version for the CP community (including young people with CP and/or their families and their carers) and
2. a version for health professionals (doctors, nurses, dentists, dietitians, physiotherapists, speech pathologists, and occupational therapists).

This Consensus Statement for the CP community aims to enable people with CP and/or their families:

- to understand and identify risk factors for respiratory illnesses in young people with CP,
- to understand ways of preventing respiratory disease in young people with CP,
- to know what assessments are recommended,
- to know what treatments are available for young people with CP with respiratory disease and why they are recommended.

The Consensus Statement for health professionals aims to help them:

- to assess risk of respiratory disease,
- to manage risk of respiratory disease,
- to make a referral for an assessment of symptoms that risk factors for respiratory disease in CP, and
- to make clinical decisions about how to care for children with respiratory disease.

CONSENSUS STATEMENT COMMITTEE MEMBERS

The members adhered to the American Academy for Cerebral Palsy and Developmental Medicine (AACPDMD) 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. **A systematic review** – which is a detailed analysis of all the research papers published 1998-2018 about treatments to prevent or manage respiratory disease in young people with CP.¹⁹ (We screened 3,347 articles and we analyzed 233 articles).
2. **A Delphi study** – which is 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.
3. **A consumer review** – In order to complete this process, we asked people with CP and their families to help us understand which treatments are acceptable and useful for preventing and managing respiratory disease in young people with CP. Carers of young people with CP notice risks to chest health and seek professional guidance about how to prevent respiratory disease. Therefore, sometimes the recommendations in this Consensus Statement are for carers or young people with CP; other times they are for health professionals. The Consensus Statement does not specify *which* health professional because: (a) that varies in different places, and (b) in most places, different health professionals work in teams.

Recommendations are classified into three types:

1. **Evidence-Based Recommendations (EBR).** These are based on the systematic review and show high or moderate support using the GRADE framework.
2. **Consensus-Based Recommendation (CBR).** These are based on agreement among the health professionals who participated in the Delphi study.
3. **Practice Points (PP).** These are important practical points that have some published evidence but participants in the Delphi study did not reach agreement about them.

LAYOUT OF THE CONSENSUS STATEMENT

The consensus statement is presented in three parts:

- PART 1:** Recognizing and managing risks to prevent respiratory disease in young people with CP;
- PART 2:** Assessing respiratory health in young people with CP;
- PART 3:** Treating and managing respiratory disease in young people with CP.

PART 1



Recognising and
managing risks to
prevent respiratory
disease in young
people with CP

To prevent respiratory disease in CP, it is important to do two things:

1. recognize risk of respiratory disease early, and
2. manage risk factors for respiratory illness.

Recent research has identified 9 risk factors.^{13-15, 22} A young person with CP who has any of these risk factors is significantly more likely to have at least one hospital admission for respiratory illness in the next 5 years.

The risk factors are:

1. Gross Motor Function Classification Scale (GMFCS - see pages 13 and 14 for more information) Level V (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. Swallowing difficulties,
4. Current seizures,
5. Frequent symptoms (daily cough or weekly sounding sound chesty or phlegmy or wheezy),
6. Gastro-esophageal reflux disease,
7. At least 2 courses of antibiotics for respiratory illnesses in past year,
8. Mealtime respiratory symptoms (gurgly voice, wheezing, coughing, sneezing, choking), and
9. Snoring every night.

These risk factors are shown in Figure 1 on page 11.

An interactive online tool to identify risk factors for respiratory disease in young people with CP can be found by typing in this address into your browser:

<https://www.telethonkids.org.au/cpchecklist>

Families of children with CP can use this online checklist to work out their risk factors.

There have been no published research trials in the past 20 years showing how to prevent respiratory disease in healthy young people with CP.²³ Therefore the following recommendations are all consensus-based recommendations (CBR).

FIGURE 1: Risk factors for respiratory hospital admissions for young people (1-26 years) with cerebral palsy.

RED FLAGS



Gross Motor Function Classification System (GMFCS) Level V
 IRR* = 23.25 (95% CI: 10.46 to 51.70)



At least one respiratory hospital admission in the last year
 IRR* = 11.8 (95% CI: 5.6 to 24.7)



At least 2 courses of antibiotics for respiratory illness in the last year
 IRR* = 5.9 (95% CI: 3.0 to 11.6)

POTENTIALLY MODIFIABLE FACTORS



Oropharyngeal dysphagia
 (requires food or drinks with modified texture OR uses a tube OR coughs and chokes on saliva)
 IRR* = 12.7 (95% CI: 7.3 to 22.1)



Frequent respiratory symptoms
 (daily cough of weekly sounding sound chesty or phlegmy or wheezy)
 IRR* = 9.4 (95% CI: 3.5 to 25.8)



Mealtime respiratory symptoms when well
 (gurgly voice, wheezing, coughing, sneezing, choking)
 IRR* = 3.8 (95% CI: 2.1 to 7.1)



Gastro-oesophageal reflux disease
 (now or previously)
 IRR* = 3.4 (95% CI: 1.8 to 6.3)



Current seizures
 (now or previously)
 IRR* = 7.6 (95% CI: 4.2 to 13.8)



Snoring every night
 (now or previously)
 IRR* = 2.8 (95% CI: 1.3 to 6.1)

* IRR (Incidence Rate Ratio) indicates the expected magnitude of the respiratory hospital admission rate of a 5-year period when the risk factor is present versus when it is absent. e.g., Young people with CP classified as GMFCS V are expected to have a respiratory hospital admission rate about 23 times greater than those classified GMFCS I to IV over a 5-year period.

Blackmore AM, Bear N, Langdon K, Moshovis L, Gibson N, Wilson AC. Respiratory hospital admissions and emergency department visits in young people with cerebral palsy: 5-year follow up. *Archives of Disease in Childhood* 2019. 745-771.

<https://adc.bmj.com/content/early/2019/06/29/archdischild-2019-317714>.

Informing parents early of risk

1.1	When a child is diagnosed with CP, professionals should discuss risk of respiratory disease with the family.	CBR
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Considering risk of respiratory disease based on child's mobility

Different children with CP have different mobility needs. Some children with CP can walk without any aids, some need the help of walking aids and others need to use a wheelchair. The grading is different for children of different ages. For more details refer to:

https://www.canchild.ca/system/tenon/assets/attachments/000/000/058/original/GMFCS-ER_English.pdf

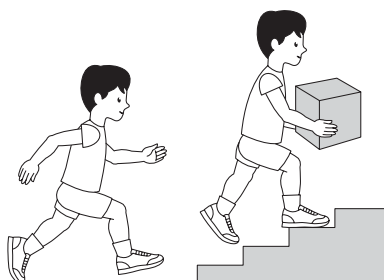
The Gross Motor Functional Classification Scale (GMFCS) classifies children into 5 levels. The five levels may be summarized as follows:

- 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 all settings and may require additional support for the head or torso

A child with a Level V GMFCS has a higher risk of respiratory disease than a child with GMFCS I, II, III, or IV.

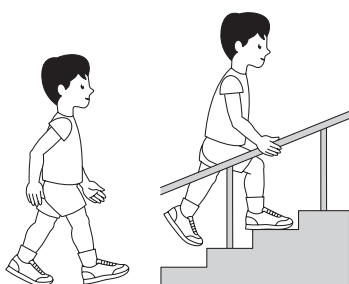
1.2	When a child is diagnosed with CP, if the child is given a GMFCS V classification, the health professional should discuss with the family the child's increased risk of respiratory disease.	CBR
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GMFCS E & R between 6th and 12th birthday: Descriptors and illustrations



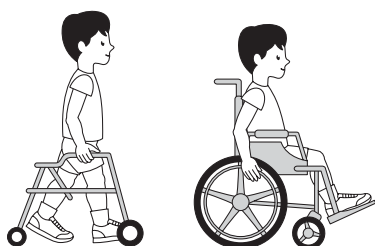
GMFCS Level I

Children walk at home, school, outdoors and in the community. They can climb stairs without the use of a railing. Children perform gross motor skills such as running and jumping, but speed, balance and coordination are limited.



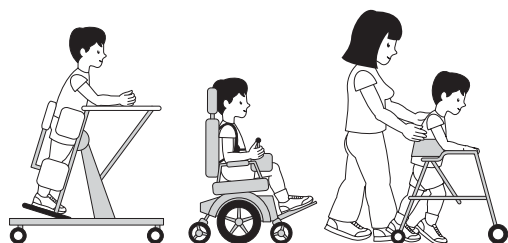
GMFCS Level II

Children walk in most settings and climb stairs holding onto a railing. They may experience difficulty walking long distances and balancing on uneven terrain, inclines, in crowded areas or confined spaces. Children may walk with physical assistance, a hand-held mobility device or used wheeled mobility over long distances. Children have only minimal ability to perform gross motor skills such as running and jumping.



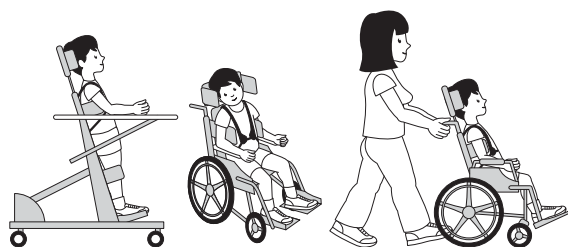
GMFCS Level III

Children walk using a hand-held mobility device in most indoor settings. They may climb stairs holding onto a railing with supervision or assistance. Children use wheeled mobility when traveling long distances and may self-propel for shorter distances.



GMFCS Level IV

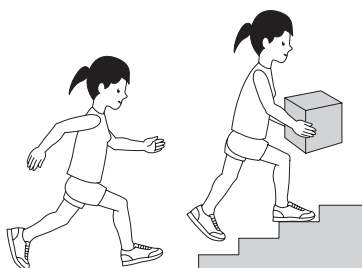
Children use methods of mobility that require physical assistance or powered mobility in most settings. They may walk for short distances at home with physical assistance or use powered mobility or a body support walker when positioned. At school, outdoors and in the community children are transported in a manual wheelchair or use powered mobility.



GMFCS Level V

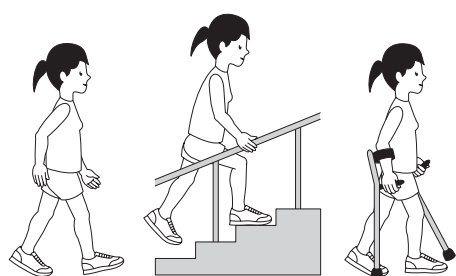
Children are transported in a manual wheelchair in all settings. Children are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements.

GMFCS E & R between 12th and 18th birthday: Descriptors and illustrations



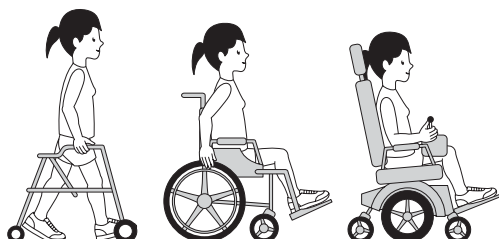
GMFCS Level I

Youth walk at home, school, outdoors and in the community. Youth are able to climb curbs and stairs without physical assistance or a railing. They perform gross motor skills such as running and jumping but speed, balance and coordination are limited.



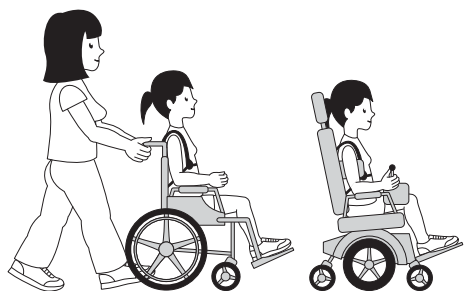
GMFCS Level II

Youth walk in most settings but environmental factors and personal choice influence mobility choices. At school or work they may require a hand held mobility device for safety and climb stairs holding onto a railing. Outdoors and in the community youth may use wheeled mobility when traveling long distances.



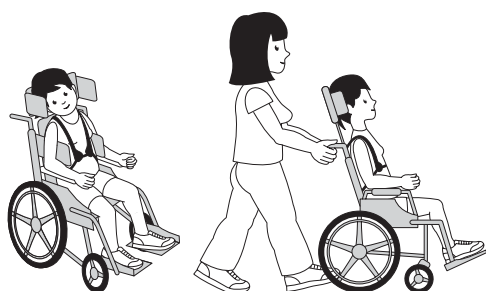
GMFCS Level III

Youth are capable of walking using a hand-held mobility device. Youth may climb stairs holding onto a railing with supervision or assistance. At school they may self-propel a manual wheelchair or use powered mobility. Outdoors and in the community youth are transported in a wheelchair or use powered mobility.



GMFCS Level IV

Youth use wheeled mobility in most settings. Physical assistance of 1-2 people is required for transfers. Indoors, youth may walk short distances with physical assistance, use wheeled mobility or a body support walker when positioned. They may operate a powered chair, otherwise are transported in a manual wheelchair.



GMFCS Level V

Youth are transported in a manual wheelchair in all settings. Youth are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements. Self-mobility is severely limited, even with the use of assistive technology.

Considering risk of respiratory disease based on swallowing abilities

Children with swallowing difficulties have a greater risk of respiratory disease than children who can eat and drink without difficulty. This is because children with swallowing difficulties may inhale food, drink, or saliva into their lungs. When food, drink, or saliva go into the lungs, bacteria may grow, and this may lead to respiratory disease.

Two assessment tools give information about a person's swallowing abilities:

1. The Dysphagia Outcome and Severity Scale (DOSS)
2. The Eating and Drinking Ability Classification System (EDACS - see page 16 for more on this)

The DOSS assesses swallowing difficulties. The DOSS classifies children into 7 levels, depending on their swallowing abilities:

- Levels 1 and 2 are unable to take nutrition by mouth.
- Levels 3, 4 and 5 are able to take nutrition by mouth, but require thickened fluids and/or thickened or puréed foods.
- Levels 6 and 7 are able to eat and drink a normal diet by mouth.

1.3	When a child is diagnosed with CP, if the child is given a DOSS classification of 1, 2, 3, 4, or 5, the professional should discuss with the family the child's increased risk of respiratory disease.	CBR
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The EDACS assesses eating and drinking abilities. The EDACS has 5 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; 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

Eating and Drinking Ability Classification System from 3 years: descriptors and illustrations



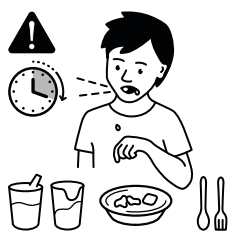
EDACS Level I - Eats and drinks safely and efficiently

- Eats a wide range of different texture foods that are age appropriate
- May be challenged by some very firm bite and chew foods
- Moves food from one side of the mouth to the other; may close lips whilst chewing
- Drinks thin or thick fluids from range of cups with consecutive swallows, including through a straw
- May cough or gag for very challenging textures
- Eats and drinks at a similar speed to peers
- Retains most food or fluid in the mouth
- Clears food from most tooth surfaces and dislodges most foods from the sides of the mouth.



EDACS Level II - Eats and drinks safely but with some limitations to efficiency

- Eats a range of food textures that are age appropriate
- Challenged by some firm bite, effortful chew, mixed and sticky textures
- Moves food slowly from one side of the mouth to the other using the tongue
- May chew with lips open
- Drinks thin or thick fluids from most cups with consecutive swallows; may drink through a straw
- Coughs or gags on new or challenging textures or when tiring
- May sometimes cough if fluid is fast flowing or large quantity taken in the mouth
- May tire if textures challenging and mealtimes will take longer than for peers
- Loses small amounts of food or fluid especially challenging textures
- Some foods will collect on some tooth surfaces and between cheeks and gums.



EDACS Level III - Eats and drinks with some limitations to safety; there may be limitations to efficiency

- Eats puree and mashed food and may bite and chew some soft chew food textures
- Challenged by large lumps, firm bite and effortful chew textures which may lead to choking and reduced efficiency
- It is challenging to move food from one side of the mouth to the other, to keep food in the mouth, and to bite and chew for safe eating
- Eating and drinking performance is variable and depends upon overall physical ability, positioning or assistance given
- May drink from an open cup but drinking from cup with a lid or spout may be required to control the flow of fluid
- May drink thickened fluids more easily than thin and may need time between sips
- May choose to drink only in certain situations such as with a trusted carer or with no distractions
- Specific food textures and positioning of food in mouth are required to reduce the risk of choking
- May cough or aspirate if fluid is fast flowing or large quantity taken in the mouth
- May tire whilst eating if food requires chewing and mealtimes will be prolonged
- Food and fluid loss is likely and food will collect on tooth surfaces, roof of the mouth and between cheeks and gums.



EDACS Level IV - Eats and drinks with significant limitations to safety

- Eats smooth purees or well mashed food
- Challenged by food that requires chewing; choking may occur if lumps are eaten
- May at times be difficult to co-ordinate swallowing and breathing when eating and drinking as shown by signs of aspiration
- It is challenging to control the movement of food and fluid in the mouth, to control mouth opening and closure, and to control swallowing, biting and chewing
- May swallow lumps whole
- May find it easier to drink thickened fluids than thin fluids; thickened fluids taken slowly and in small quantities from an open cup may increase control whilst drinking
- May choose not to drink fluids or to drink only in certain situations such as with trusted carer
- Likely to need time between mouthfuls to swallow repeatedly before continuing
- Will require specific food textures, fluid consistency, techniques, skilled carers, positioning and modified environment to reduce risks of aspiration and choking and increase efficiency
- May tire whilst eating and mealtimes are likely to be prolonged
- Significant food and fluid loss from the mouth
- Food may become stuck on tooth surfaces, roof of the mouth and between teeth and gums
- Supplementary tube feeding may be considered.



EDACS Level V - Unable to eat or drink safely – tube feeding may be considered to provide nutrition

- May manage very small tastes or flavours
- Ability to manage small tastes and flavours will be affected by positioning, personal factors and environmental features
- Unable to swallow food or drink safely due to limitations to the range and co-ordination of movement for swallowing and breathing
- It is likely to be challenging to control mouth opening and tongue movement
- Aspiration and choking are very likely
- Harm from aspiration is evident
- May require suction or medication to keep airway clear of secretions
- Alternative means of providing nutrition such as tube feeding may be considered.

The EDACS can be used to assess children only after the age of 3 years.

1.4	When a child is diagnosed with CP, if the child is given an EDACS classification of III, IV, or V, the professional should discuss with the family the child's increased risk of respiratory disease.	CBR
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Identifying and preventing aspiration

Everyone swallows many hundreds of times every day. Most of the day, people only swallow their saliva. During meals, they need to swallow much more, and that is when they are most likely to notice whether they have swallowing difficulties.

When a person swallows foods, drinks, and saliva, these substances should go down the foodpipe into the stomach. Sometimes foods, drinks, saliva, or vomit accidentally go down the windpipe into the lungs. In other words, it “goes down the wrong way”. This is called **aspiration**. Sometimes the person will cough or choke, but most of the time there are no outward signs of aspiration and so it is called “silent aspiration”. Silent aspiration can be detected using a videofluoroscopy (a moving X-ray taken during swallowing).

Aspiration can cause respiratory disease. This happens because food, drink, saliva, or vomit goes into the lungs. This can cause bacteria to grow, and leading to respiratory disease.

1.5 (i)	Health professionals should identify aspiration in young people with CP as early as possible.	CBR
1.5 (ii)	Health professionals should assess young people with CP to identify risk factors for aspiration as early as possible.	CBR
1.5 (iii)	Health professionals should treat risk factors for aspiration as early as possible.	CBR

There are 4 risk factors for aspiration.

1. Swallowing difficulties
2. Uncontrolled seizures
3. Reflux
4. Drooling saliva (which is a marker of swallowing difficulties)

The recommendations for each of these are as follows:

Swallowing difficulties

Health professionals should monitor young people with CP who have swallowing difficulties to see whether they are aspirating saliva, food, and/or drinks. Note that aspiration may involve gagging, choking or coughing or it may be silent.

1.6	If young people with CP are suspected of aspirating, health professionals and families should consider a comprehensive assessment by a multidisciplinary team (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, health professionals and families should consider ways of training the muscles of the mouth.	CBR
1.9	If young people with CP have trouble controlling their saliva, health professionals should consider the way that they are seated, the position of their head, and other ways of preventing them from aspirating their saliva.	CBR
1.10	If young people with CP are aspirating, health professionals should review them regularly	CBR

Uncontrolled seizures

Some young people with CP have seizures. During a seizure, a person might become confused, have repetitive or jerky movements that they cannot control, or lose consciousness. They may vomit, drool or stop breathing. Any vomit or excess saliva can be aspirated into the lungs. Seizures can often be controlled by medications, but sometimes medications do not fully control the seizures. Sometimes, medications cause sedation which leads to increased drooling and further aspiration.

1.11	Health professionals should monitor young people with CP for ongoing seizures.	CBR
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1.12	When seizures become uncontrolled, health professionals should refer young people with CP to a neurologist.	CBR
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Reflux

Reflux occurs when the contents of the stomach come up the foodpipe. Sometimes this can go into the windpipe and into the lungs. This can cause bacteria to grow in the lungs, leading to respiratory disease.

1.13	Health professionals should assess and manage reflux.	CBR
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Drooling saliva

Two in five children with CP over the age of 4 years drool with their saliva.⁷ This is not because they produce too much saliva, but because they have trouble managing their saliva.²⁴ Sometimes, saliva collects in large amounts at the back of the throat, where it is aspirated into the lungs and leads to ongoing respiratory illnesses.^{25, 26}

1.14	Health professionals 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 Toxin, or salivary gland surgery to manage drooling.	CBR

Optimising airway clearance

Most people keep their airways clear when they are healthy by breathing deeply, sighing, taking bigger breaths when active, changing positions regularly, and coughing. Coughing clears secretions from the airways. However, some young people with CP are not able to clear their airways efficiently because their cough is too weak.

Physiotherapists can show carers ways to clear the airway secretions of young people with CP. These ways include manually assisted cough and suctioning.

1.17	Physiotherapists should show carers techniques to maintain clear airways.	CBR
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1.18	Health professionals should identify young people with CP who have an ineffective cough and teach families and caregivers techniques to optimize cough to manage secretions. (Refer to Treatment section 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, health professionals should consider prescribing regular chest physiotherapy. (Refer to Treatment section 3.9-3.18 for additional recommendations relating to this.)	CBR
1.20	If young people with CP have upper airway obstruction, health professionals manage it, where possible, using a change in position and tone management.	CBR
1.21	Health professionals and carers help young people with CP to sit upright or lie in positions that improve airflow through the lungs and expansion of the ribcage.	CBR
1.22	If young people with CP have a history of stopping breathing for more than 10-20 seconds, snoring, or if they have large tonsils, health professionals 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, physiotherapists should use preventative measures, such as airway clearance regimes. (see Treatment section 3.19-3.20)	CBR
1.24	Health professionals should show young people with their families how to optimize movement of the ribcage to prevent problems that arise when the lungs cannot expand fully (i.e. restrictive lung disease). This includes: <ul style="list-style-type: none"> a. physiotherapists showing carers how to maintain chest movement, b. maximizing physical activity and minimizing sitting and lying (e.g., by regular position changes for young people with CP who cannot walk), c. managing movement disorders, d. health professionals assessing young people with CP for scoliosis (curvature of the spine) and preventing or managing it, and e. where surgery for scoliosis is being considered, a multidisciplinary team evaluating risks and benefits. 	CBR

Optimising nutrition

There is no research evidence to show whether improving nutritional intake in young people with CP prevents respiratory disease. However, in people without CP, there is evidence that insufficient nutrition leads to weakened immune responses.²⁷⁻³⁰ In children without CP, insufficient nutrition may interfere with the growth of the lungs, diaphragm, and other muscles needed for breathing, so that children who do not take enough nutrition have less strength and lower resistance.³⁰ When people have respiratory infections, they need more energy to breathe and to fight the infection. On the other hand, infections reduce the appetite. This means that people who are underweight may struggle to find enough energy stores to manage the infection.^{28, 30} Therefore, there is consensus agreement about the following recommendations.

1.25	If young people with CP have poor nutritional status, dietitians should optimize their nutritional intake.	CBR
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Optimising 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.³¹ Many studies show that the more physical activity, the better the lung function in children and adolescents without disabilities.^{32,33}

Studies of swimming,³⁴ breathing exercises,³⁵ arm exercises with elastic bands,³⁶ and feedback respiratory training plus movements,³⁷ all reported improvements in lung function in healthy children with CP. There is expert consensus that lung function, physical activity, and fitness should be optimized in young people with CP.

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

Maintaining dental hygiene

The mouth can be a reservoir for the bacteria that cause respiratory illnesses,^{38,39} Among people with no CP, those with poor oral health are more likely to develop pneumonia, whereas improving oral health reduces the risk of pneumonia.^{37,40} This is likely to be true for people with CP as well. There are particular challenges to young people with CP maintaining good oral health. Some young people with CP cannot hold their mouths open and steady for long enough for a dental examination or even for the teeth to be cleaned properly. These challenges may increase their risk of poor oral health and respiratory illnesses.

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

Influenza can lead to serious respiratory illnesses, especially in children who are vulnerable. It is very common and easy to catch. The best way to prevent flu is to have an annual vaccination. It is more effective to vaccinate the whole family of a young person with CP than just the young person alone.

1.29	Young people with CP and their families should be vaccinated annually against influenza.	CBR
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Reducing exposure to tobacco smoke

The effects of smoking on lung function and chest health are well known. A person doesn't need to smoke cigarettes to suffer the ill effects of tobacco smoking. Breathing in other people's tobacco smoke (passive smoking) has similar effects to smoking. Numerous studies show that people who are exposed to passive smoke have more respiratory illnesses.⁴¹⁻⁴⁵

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

People with asthma have difficulty breathing in and out at times because their airways become narrow, the airway muscles get tighter, and the airway walls become swollen. Asthma is usually controlled by medications.

1.31	Health professionals should be alert for asthma in young people with CP.	PP
1.32	Doctors should assess the way that young people with CP respond to asthma medications. If symptoms do not improve, then doctors should consider stopping the medication.	PP



Ongoing screening 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.

Some young people with CP are at higher risk than others, and these need to be screened for risk factors more often.

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: <ul style="list-style-type: none">• A hospital admission for a respiratory illness in the past 12 months.• GMFCS Level V.• EDACS Levels III, IV or V and/or (for children under 3 years of age) DOSS Levels 1, 2, 3, 4, or 5.	CBR
1.34	Health professionals should consider screening young people with CP for risk of respiratory disease more often if they meet any of the following criteria: <ul style="list-style-type: none">• An increase in hospital admissions for respiratory illnesses.• A hospital admission for a respiratory illness since the last review.• Poor control of seizures.• Unexplained weight loss or gain.• A change from taking food and drink by mouth to taking nutrition by tube.• Evidence of aspiration. (Evidence may be from a clinical swallow assessment, choking episodes, chest X-ray, or videofluoroscopy.)• Deterioration in the ability to sit, stand, or walk, especially if sitting or standing become worse.• Deterioration in control of the mouth and swallowing muscles and difficulty managing secretions.• Scoliosis.• Changes in general health that might affect the ability to swallow and cough (e.g., pain, fatigue).• Any other concern related to risk of respiratory disease that may be identified by family or health professionals.	CBR

PART 2



Assessing respiratory health in young people with CP

Initial assessments

Health professionals should assess young people with CP for their risk of respiratory disease. The assessment includes:

- a detailed interview of the young person with CP and/or their family about their previous health specifically focused on factors such as breathing, coughing, sleeping and chest related hospital admissions and
- examining the young person with CP directly.

This assessment will reveal any concerns of the young person with CP or the family. The assessment will also identify the factors that might put a young person with CP is at risk of respiratory disease. Recent changes in the ability to sit, stand, walk, swallow, eat or drink should be noted. The GMFCS level, EDACS level, and DOSS level should also be noted.

2.1	Health professionals from different disciplines (e.g., speech pathologist, physiotherapist, pediatrician) should do different parts of the assessments.	CBR
2.2	Health professionals should assess symptoms of respiratory disease.	CBR
2.3	Health professionals should interview the young person with CP and/or their family about their previous health and treatments as far as they relate to the risk of respiratory disease. This assessment should include the following: <ol style="list-style-type: none">a. GMFCS,b. EDACS or DOSS by a speech pathologist if under 3 years of age,c. Seizures,d. Reflux,e. Drooling with saliva, andf. Episodes of aspiration, wheezing or noisy breathing.	CBR

2.4	<p>Health professionals should interview the young person with CP and/or their family about their respiratory illnesses during the last 12 months. This assessment should include questions about:</p> <ul style="list-style-type: none"> a. use of antibiotics (how often? what type? for how long?), b. hospital admissions for respiratory illnesses, c. hospital admissions where the young person with CP developed a respiratory illness after being admitted to hospital, and d. non-invasive ventilation (breathing support from a machine using a mask). 	CBR
2.5	<p>Health professionals should give a thorough respiratory examination to the young person with CP. (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.)</p>	CBR
2.6	<p>Health professionals should assess how well the young person with CP is able to manage secretions from the airways. This should be done when the young person is well and again when the young person is unwell.</p>	CBR
2.7	<p>Health professionals should assess nutritional status (weight, height, rate of growth, blood test, dietitian review).</p>	CBR
2.8	<p>Before changing the way that a young person with CP takes food and drink (e.g., using thickened textures or a tube), health professionals should consider whether the health of the young person with CP is stable and well.</p>	CBR
2.9	<p>Health professionals from different disciplines (e.g., speech pathologists, occupational therapists, physiotherapists, and dietitians) should assess the eating and drinking of a young person with CP and whether the young person is at risk of aspirating food, drink, saliva, or vomit. This should be done when the young person is well and again when the young person is unwell.</p> <ul style="list-style-type: none"> a. Where swallowing difficulties are suspected, speech pathologists should assess oropharyngeal motor dysfunction. b. Swallowing using a range of different food textures and different drink textures should be assessed. 	CBR

2.10	Health professionals should assess young people with CP for scoliosis.	CBR
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Referrals for diagnostic tests

When the assessments above have been completed, health professionals may refer a young person with CP for further tests:

2.11	A videofluoroscopy should be performed on young people with CP when the health professionals' assessments show that there may be a risk of aspiration.	CBR
2.12	A sleep study (showing overnight blood oxygen levels) should be considered for young people with CP with symptoms of airway obstruction or apnea.	CBR
2.13	Sputum culture, chest x-ray and a chest CT (more detailed x-ray images of the chest and lungs) may provide additional information to guide treatments in some instances.	PP



PART 3



Treating and
managing respiratory
illness in young people
with CP

Treatment of respiratory disease, 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.

Recommendations for prevention included avoiding tobacco smoke, maintaining flu vaccinations, and ensuring good dental care. All of these apply to treatment of respiratory disease too.

The following recommendations are for treatment of young people with CP with respiratory illnesses.

General recommendations regarding management of respiratory illness

For young people with CP with respiratory illness, it is recommended that:

3.1	All the recommendations from <i>Part 1: Recognizing and Managing Risks to Prevent Respiratory Illness in Young People with CP</i> apply to treatment of respiratory illness too.	CBR
3.2	Health professionals should consider assessing and treating any signs of respiratory illness as soon as possible	CBR
3.3	Teams of health professionals from different disciplines (e.g., doctors, nurses, physiotherapists, speech pathologists) should work together to manage respiratory illnesses in young people with CP including allocating a team leader who takes responsibility to coordinate care decisions with the young person with CP and their family.	CBR
3.4	When a young person with CP has a respiratory illness, health professionals should investigate the underlying causes and guide treatments differently, depending on the cause.	CBR

Use of medications

Medications to manage respiratory illnesses may include:

- antibiotics to treat bacterial infections,
- bronchodilators to open the airways,
- anti-inflammatory agents to manage spasms of the airways, and
- mucolytics to help thin secretions,
- botulinum toxin to reduce drooling with saliva.

3.5	As soon as a young person with CP gets a respiratory illness in the lungs, health professionals should treat it with antibiotics using antibiotic guidelines.	CBR
3.6	Health professionals should manage drooling with saliva in young people with CP with respiratory illnesses. Health professionals should bear in mind that interventions that thicken airway secretions may cause the airways secretions to form a plug blocking one of the airways.	CBR

There is published evidence that salivary gland botulinum toxin⁴⁶⁻⁴⁹ and salivary gland surgery reduce respiratory infections.⁵⁰⁻⁵² However, in all these studies, the authors did not compare the group of children who had treatment compared to the group of children who did not receive the treatments. (So the evidence is not very strong).

There is evidence from published case studies of two children that nebulized tobramycin may reduce respiratory hospital admissions and days for pneumonia.⁵³

There was no evidence identified in the systematic review and no consensus regarding the use of mucolytic agents, that is, medications that make airway secretions thinner.

Optimising general health

3.7	When young people with CP have respiratory illnesses, health professionals should consider whether they are taking in enough nutrition to enable them to recover from an infection.	CBR
3.8	When young people with CP have respiratory illnesses, health professionals should routinely assess and manage reflux.	CBR

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

There is published evidence that respiratory illnesses are improved by gastrostomy (surgical insertion of a tube so that food can be supplied directly into the stomach) sometimes with fundoplication (surgery to prevent reflux).⁵⁴⁻⁵⁷ One study reported no difference.⁵⁸ All of these studies had only one group of children; they did not compare the children who had the treatments with similar children who did not. Several of these studies also reported significant negative outcomes following these treatments. However, many of these outcomes were not necessarily caused by the treatments: children who have these treatments usually have very complex conditions and are at risk of developing other problems when they are sick.

Optimising airways

There is published evidence that surgery to clear airway obstructions in young people with CP improves blood oxygen levels,^{59, 60} and improves sleep and the ability to function during the daytime.⁶¹ However, none of these studies compared the children who had airway obstruction surgery with children who had no surgery. The following recommendations are for treatment of young people with CP with respiratory illnesses or with difficulties clearing the airways of secretions. There was no consensus reached on the use of Non-Invasive Ventilation (NIV) for the treatment of upper airway obstruction.

Physiotherapy for airway clearance

There are not many studies published for breathing physiotherapy treatments in young people with CP. It is therefore important that when physiotherapists prescribe physiotherapy for the airways that they monitor and teach parents and carers how to monitor the young person's response to the treatments carefully. This includes monitoring for both positive responses, but also intolerance to the treatments.

3.9	When young people with CP have respiratory disease, or difficulties getting rid of mucus, health professionals should consider referring them to a physiotherapist with experience treating breathing difficulties.	CBR
3.10	When young people with CP have respiratory illnesses, physiotherapists should prescribe chest physiotherapy, and educate carers on changes to routine positioning to improve breathing and lung expansion.	CBR
3.11	When young people with CP have respiratory illnesses, physiotherapists should assess whether the cough is strong enough to keep the airways clear of secretions.	CBR
3.12	When young people with CP have respiratory illnesses, physiotherapists should show carers how to position them for 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 a weak cough but safe swallowing, physiotherapists should show carers on how to assist their cough manually.	CBR
3.14	When young people with CP have respiratory illnesses, if their cough is not strong enough to clear airway secretions from their airways and/or they cannot swallow safely, health professionals should consider showing families how to suction.	CBR
3.15	When young people with CP have respiratory illnesses, if they are able to cough up and clear their airway secretions safely, physiotherapists should prescribe ways of keeping the airways clear.	CBR

3.16	When young people with CP have a wet cough that continues for at least 4 weeks, physiotherapists should prescribe long-term ways of keeping the airways clear, and these should be done every day.	CBR
3.17	When young people with CP have respiratory illnesses, physiotherapists should monitor ways of keeping airways clear, and change them as needed.	CBR
3.18	When young people with CP have respiratory illnesses and are using ways of keeping their airways clear, physiotherapists should monitor them for signs of intolerance.	CBR

Although there was consensus about positioning to optimise lung function, there is no research evidence for the effectiveness of this intervention for respiratory health.^{62, 63}

There was no consensus reached on the use of mechanical assistive devices, such as mechanical insufflation-exsufflation or high frequency chest wall oscillation to help remove mucus or secretions for young people with CP with respiratory illnesses. There is some evidence that high frequency chest wall oscillation reduces respiratory hospital days, pneumonia episodes and days on antibiotics,⁶⁴⁻⁶⁷ but these studies had no comparison group of young people with CP who did not receive the treatments, so the evidence is not very strong. One well-designed study of high frequency chest wall compression has been published. This study did not find any evidence of an effect on hospital days or antibiotics, but that may have been because the study had only a small number of children (only 9 of the participants were young people with CP).⁶⁸ There is no current evidence that mechanical insufflation-exsufflation (also known as “Cough Assist Machines”) makes a difference to respiratory health in young people with CP.⁶⁹

Reviewing mealtime management plans

3.19	When young people with CP have respiratory illnesses or any other illnesses, speech pathologists should assess their swallowing and work with the young person and family to create a management plan to ensure that eating and drinking are safe and the young person can get as much nutrition as they need.	CBR
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3.20 When young people with CP have respiratory illnesses, and a swallowing assessment shows that tube feeding may be beneficial, this should be discussed with the young person with CP, family, medical team, and dietitian.

CBR

There is published evidence that providing mealtime management advice to mothers of children with CP reduces respiratory illnesses (although this study did not compare these parents with parents who did not receive advice).^{70, 71}

There is also published evidence that children who had treatment guided by a videofluoroscopy improved in their respiratory health (but there was no comparison with children who did not have a videofluoroscopy).⁷²

In children who take nutrition by tube, there is published evidence that thickening the formula reduces their reflux, cough, and wheeze. The children's reflux, coughing and wheezing were compared with and without the thickened formula.⁷³



ADVANCED CARE PLANNING

Discussions with parents about the death of a child are not easy to have. However, the possibility of an early death from a respiratory illness for a child with CP within GMFCS level V may become a reality. The exact timing of death is difficult to predict. Often a sign of deterioration can be an increasing number of hospital admissions. Advanced Care Planning is the term used for the discussion between parents (and when appropriate the individual with CP) about treatment options, given that death is a possibility. Ideally, doctors and families make decisions together about treatments. Decisions may range from “full resuscitation”, meaning that every available treatment is offered to increase length of life, through to treatments that focus on reducing pain and improving comfort, knowing that the individual may not survive. Family preferences and decisions can be written in the medical records, which may assist doctors during unplanned hospital admissions. Even if no decisions are reached, it is helpful for other doctors to understand the family’s beliefs and plans.

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.

The strengths of this Consensus Statement are:

- It is targeted specifically at young people with CP.
- 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.
- It is guided by consumer input.
- It takes into account specific individual risk factors.

The major limitation of this Consensus Statement is that it lacks a strong evidence base.

Each young person with CP will respond differently to the treatments described in this 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.

FUTURE RESEARCH

The systematic review showed that there was not very much 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 patient experience 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. Our systematic review and Delphi study both showed that health professionals and researchers believe a lot of different kinds of treatments may help young people with CP with respiratory disease. But there is very little evidence for any of these in CP (although many of these treatments have been used successfully with people who do not have CP).

Treatments that need further research are:

- influenza vaccination,
- preventative or early use of antibiotics,
- conventional physiotherapy techniques,
- airway clearance techniques,
- exercise,
- positioning,
- non-invasive ventilation,
- dental health,
- pharmaceutical and surgical methods of reducing drooling with saliva,
- surgical management of upper airways,
- mealtime management,
- training of mouth muscles,
- electrical stimulation of swallowing muscles,
- nutritional management,
- management of reflux,
- gastro-intestinal surgery,
- spinal surgery, and
- management of seizures.

At the same time, research is also needed to understand what causes some young people with CP to get repeated respiratory illnesses and how the disease progresses over time.

All published evidence is about single treatments for respiratory disease in young people with CP. There is no published evidence on multiple treatments for respiratory disease in young people with CP. However, multiple treatments are often 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,^{14,15,76} it is recommended that preventative interventions for these children be explored.

All future research on respiratory disease in CP needs to be well designed, high quality, with sufficient numbers of young people with CP in the trials, and sufficient follow-up periods to find out whether treatments are effective. Future research also needs to include patient-reported outcomes (e.g., activities, participation, and quality of life).

DISCLAIMER

This consensus is based on a systematic review of the current published literature published in 2019,¹⁹ 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 five years from the date of publication in order to assess uptake and impact of the recommendations, and to review new knowledge that may affect the recommendations.

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THIS CONSENSUS STATEMENT IS A COLLABORATION BETWEEN



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Child and Adolescent Health Service



Perth Children's Hospital



The Sydney
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