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# RESPIRATORY HEALTH IN CEREBRAL PALSY

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## DEFINITIONS

Individuals with cerebral palsy (CP) are susceptible to respiratory illnesses such as upper and lower respiratory tract infections, the latter including pneumonia which is commonly due to aspiration. Symptoms of illness may be fever, lethargy, cough, sore throat, runny nose, increased secretions, gurgly breathing or shortness of breath. Oropharyngeal dysphagia (OPD) is the medical term for difficulty swallowing. The motor control issues in CP which cause the physical disability can impact on all stages of swallowing, from closure of lips and formation of a bolus, to the pharynx, larynx and beyond. Oropharyngeal dysphagia or reflux may cause aspiration pneumonia. Poor postural control and subsequent skeletal deformity leads to restrictive lung disease, which is respiratory disease from impaired elasticity of the lungs and mechanics of the chest wall, reducing lung capacity. This contributes to impaired respiratory reserve. A weak cough may cause mucous retention, leading to atelectasis, chronic inflammation, and infection. This may lead to suppurative lung disease, bronchiectasis, and obstructive lung disease.

Issues of respiratory health in CP are interconnected, multifactorial and multisystemic. Treatments may be in the home or require a hospital admission.

## WHY IS RESPIRATORY HEALTH IN CEREBRAL PALSY IMPORTANT?:

- The leading cause of death in children and young people with CP is respiratory illness.
- For adults with CP, death from respiratory illness is 14 times higher than for their peers with no disability.
- For children and young people with CP, respiratory illness is the most common cause of presentation to the emergency department and is the most common cause of prolonged hospital admissions.
- Following an initial respiratory-related admission, the respiratory re-admission rate over the next year is 70%.
- Risk factors can be addressed to minimize aspiration which may help prevent further respiratory illness.
- Treatment of infection should be managed without delay.
- The goal of prevention and treatment is to improve longevity and quality of life for individuals with CP.

## TARGET POPULATION

The risk of respiratory illness should be assessed in all individuals with CP. The strongest predictor for respiratory illness is being classified within Gross Motor Function Classification System (GMFCS) Level V. (This level of GMFCS means any person with CP who uses a wheelchair and is unable to maintain their head or trunk position without adaptive equipment.) Also at high risk is an individual with CP who has had a respiratory admission in the past year, or has been treated with two or more courses of antibiotics for chest infections in the past year. This pathway is for those identified as individuals at high risk of respiratory illness.

## TARGET CLINICAL PROVIDERS

Physicians/Nurses/Therapists caring for individuals with CP.

## ASSESSMENT

The severity of the respiratory issues for each child will depend on individual factors. However, evidence-based red flags for individuals with CP are: having CP and being classified within GMFCS Level V; having had a respiratory admission in the past year; or having been treated with two or more courses of antibiotics for chest infections in the past year. Important and potentially modifiable risk factors for respiratory illness include: OPD in those < 3 years of age on the Dysphagia Outcome Severity Scale (DOSS) Levels 1-5; or OPD in those > 3 years of age on the Eating and Drinking Ability Classification System (EDACS) Level III-V. (These scores highlight that the presence of any OPD is important.) Other modifiable risk factors include: mealtime respiratory symptoms (gurgly voice, wheezing, coughing, sneezing, choking); frequent respiratory symptoms (daily cough or weekly sounding chesty, phlegmy or wheezy); present or past gastro-esophageal reflux disease (GERD); night-time snoring every night; and uncontrolled epilepsy. Clinicians should identify and manage these risk factors.

A thorough assessment with a respiratory history and physical examination is required. A comprehensive history should include gaining an understanding of concerns, care, comfort and goals.



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A detailed screen for red flags and potentially modifiable risk factors should be completed. An understanding of swallowing abilities and management of secretions on well and unwell days should be obtained. The physical examination should include the respiratory and heart rate, oxygen saturation level, the pattern of chest wall movement, work of breathing, chest wall shape, palpation and auscultation. Tonsils and turbinates should be visualized. Strength and efficiency of cough should be assessed. Nutritional status should be assessed. Skeletal deformities such as kyphosis and scoliosis should be recorded. Surveillance should be at least annual, with a view to increasing surveillance with any changes in clinical status. Changes in clinical status include: increased respiratory admissions since last review; any respiratory-related concern by family/carers/individual with CP or clinician; evidence of aspiration or change in clinical state affecting ability to manage airway clearance; worsening OPD; difficulties managing secretions or feeds; deterioration in motor skills; or presence of skeletal deformities.

## MANAGEMENT

**Recognize risk, discuss early and prevent recurrence**

- Early discussion about the impact that respiratory health has on morbidity and mortality with parents/carers of children with CP. This is important for those at high risk; namely those with CP classified within GMFCS Level V (or at high-risk of CP) and those with OPD.
- Consider annual surveillance and interdisciplinary assessment and the need for further investigations, referrals and reviews.
- Consider referral to respiratory physician.

**Manage risk factors for aspiration**

- Optimize gastro-esophageal reflux disease (GERD), seizure control and drooling. (Drooling may be a marker of OPD and management is described in AACPDM Sialorrhea in Cerebral Palsy Care Pathway.)
- Minimize Aspiration
  - A clinical assessment of swallowing by a speech pathologist (or occupational therapist).
  - Mealtime management prescription includes safe swallow strategies, mealtime positioning and equipment, level of assistance, risk assessment, quality of life needs and informed choice. Mealtime management strategies aim to prevent or reduce cough, wheeze, aspiration and choking.
    - A video fluoroscopy swallow study is indicated if ongoing aspiration is suspected, if there are respiratory symptoms or if there is clinical complexity.
  - A plan for well and unwell days should be made.

**Optimize airway clearance**

- Physiotherapist input is recommended for assessment and for prescription, titration and safety of a targeted airway clearance therapy when this is indicated. This includes provision of education to the individual, carer and parents. Suction may be an important adjunct but cease airway clearance therapy if there is risk of aspiration, especially in those with an ineffective cough, respiratory muscle weakness or GERD.
- Assess upper airway. Upper airway obstructive symptoms when awake may be remediable with reduction of tone medications (which may have exacerbated an iatrogenic hypotonia). Reversibility of upper airway obstruction may be assessed by an ear, nose and throat surgeon. Snoring or obstructive sleep apnea may be treated with simpler surgical interventions such as adenoidectomy. Other causes of multilevel obstruction, especially if there are night and day symptoms, require individualized decision making with a multidisciplinary team.

**Optimize chest wall mobility**

- Physiotherapist and inter-disciplinary input are recommended for assessment, prescription and optimization of chest wall mobility and posture.

**Optimize general health**

- For example: maintain fitness and physical activities to improve lung health as well as muscle strength, optimize nutritional status and ensure good dental hygiene, keep immunizations up to date, avoid tobacco smoke, and manage concomitant asthma.

## TREATMENT FOR INDIVIDUALS WITH PNEUMONIA

- Consider early investigation and treatment of most likely pathogen based on local guidelines.
- Consider airway clearance therapy and suction.

## TREATMENT FOR INDIVIDUALS WITH ESTABLISHED RESPIRATORY ILLNESS

- Continue with prevention strategies and regular assessment.
- Continue annual clinical review of the individual focusing on the respiratory system, red flags and modifiable risk factors. Increase surveillance with any changes in clinical status.
- Focus on airway clearance therapy and assess its effectiveness, tolerability and safety.



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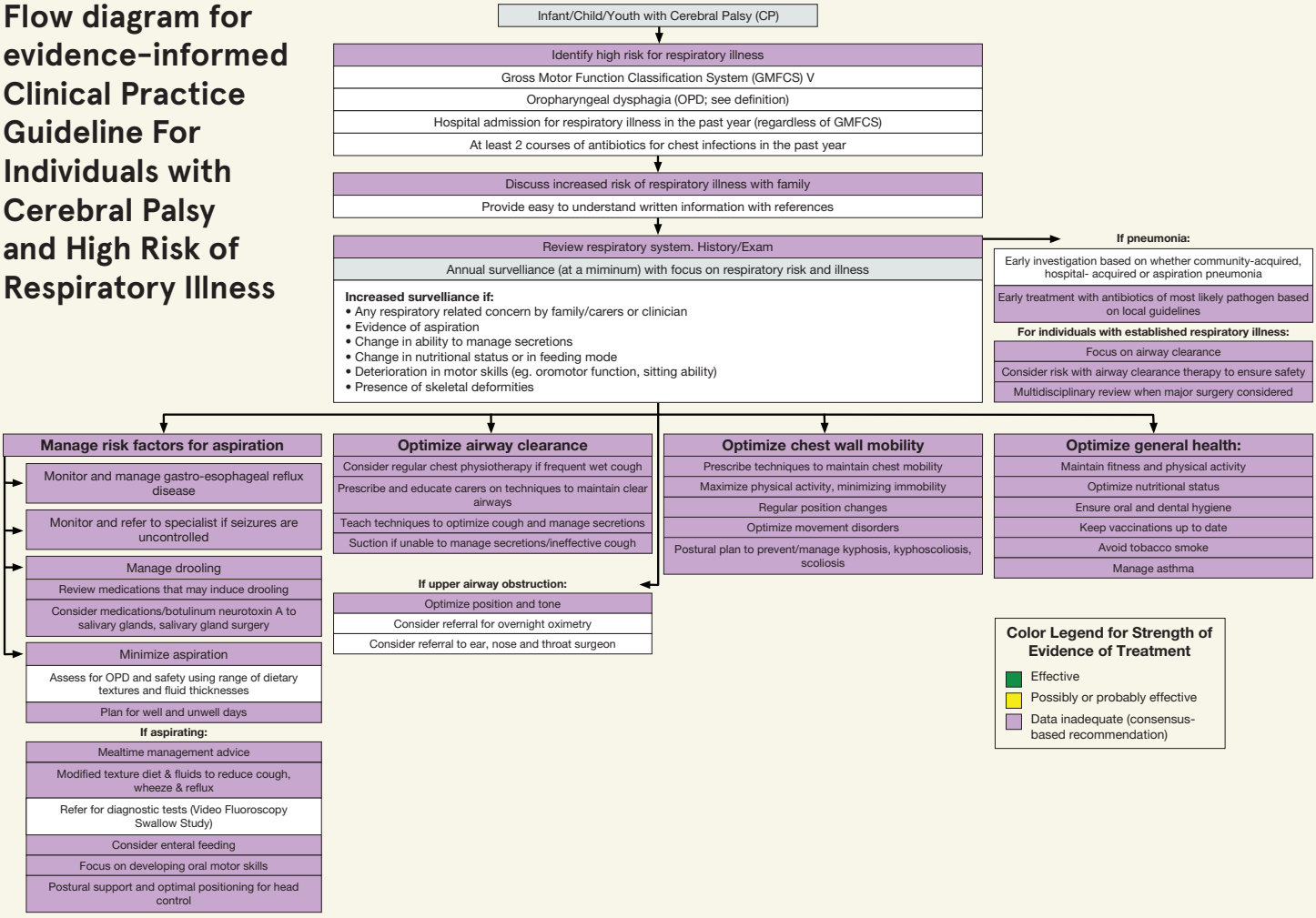
· For those with recurrent respiratory infections/established respiratory disease, ideally a shared decision model with an interdisciplinary team, primary pediatrician, family/carers and patient is the goal. The team may need to be broadened to include respiratory physicians, surgeons, anesthesiologists, physiatrists or palliative care physicians as needed.
Decisions about interventions should take into account the condition and quality of life of the patient. This does not preclude full intensive care or major surgery, if the balance of risk and benefit is agreed to be appropriate.

## Flow diagram for evidence-informed Clinical Practice Guideline For Individuals with Cerebral Palsy and High Risk of Respiratory Illness




*The purpose of this document is to provide health care professionals with key facts and recommendations for the assessment and treatment of respiratory health in children and youth with cerebral palsy. This summary was produced by M.S. Cooper (team lead for Care Pathway), A.M. Blackmore, N Gibson, A Chang, L Culloton, J Depiazzi, J Gains, A Jaffe, W Kong, K Langdon, R Marpole, L Moshovis, K Pavleski, A.C. Wilson. Consulting group: G Antolovich, G Baikie, J Chiang, C Fairhurst, AR Harvey, S Louey, P McNamara, M Proesmans, D Reddihough, L Robson, R Ross Russell, N Wimalasundera. The summary is based on a systematic review by 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 Sep;45(5):754-771. doi: 10.1111/cch.12703. Epub 2019 Jul 30. PMID: 31276598. We did not identify any interventions aimed at preventing respiratory illness in this population. The following algorithm was formulated by consensus-based recommendations: Gibson N, Blackmore AM, Chang AB, Cooper MS, Jaffe A, Kong WR, Langdon K, Moshovis L, Pavleski K, Wilson AC. Prevention and management of respiratory disease in young people with cerebral palsy: consensus statement. Dev Med Child Neurol. 2021 Feb;63(2):172-182. doi: 10.1111/dmcn.14640. Epub 2020 Aug 9. PMID: 32803795; PMCID: PMC7818421.The consensus-based recommendations included recognizing those at risk and managing risks to prevent respiratory illness. However, health care professionals should continue to use their own judgement and take into account additional relevant factors and context. The AACPDM is not liable for any damages, claims, liabilities, or costs arising from the use of these recommendations including loss or damages arising from any claims made by a third party.*

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