BTS Clinical Statement:
Community acquired pneumonia in people with learning disability

Draft 11 April 2022

Available for public consultation from
11 April 2022 to 13 June 2022

Contact:
British Thoracic Society,
17 Doughty St, London WC1N 2PL

miguel.souto@brit-thoracic.org.uk

A response form is available on the BTS website.
Please send your responses to Miguel Souto by 5pm on Monday 13th June 2022

BTS Clinical Statement:
Community acquired pneumonia in people with learning disability

Julian Legg[1,2], Jamie-Leigh Allen[1], Morag Andrew[3], Charlotte Annesley[4], Michelle Chatwin[5,6], Hannah Crawford[7,8,9], Joanna Elverson[10,11], Julian Forton[12,13], Kate Oulton[14], Kate Renton[1,15], Alison Tavare[16], Hilary Tedd[10], A John Simpson[10,17]

1. Southampton Children's Hospital, University Hospital Southampton NHS Foundation Trust, Southampton, UK
2. University of Southampton, Southampton, UK
3. Great North Children's Hospital, Newcastle upon Tyne NHS Hospitals NHS Foundation Trust, Newcastle upon Tyne, UK
4. North Middlesex University Hospital NHS Trust, London, UK
5. Clinical and Academic Department of Sleep and Breathing, Royal Brompton Hospital, Guys and St Thomas' NHS Foundation Trust.
6. NMCCC, National Hospital for Neurology and Neurosurgery, University College London NHS Foundation Trust
7. Tees, Esk and Wear Valleys NHS Foundation Trust, Darlington, UK
8. Teesside University, Middlesbrough, UK
9. University of Central Lancashire, Preston, UK
10. Newcastle upon Tyne NHS Hospitals NHS Foundation Trust, Newcastle upon Tyne, UK
11. St Oswald's Hospice, Newcastle upon Tyne, UK
12. Noah's Ark Children's Hospital for Wales, Cardiff, UK
13. Cardiff University, Cardiff, UK
14. Centre for Outcomes and Experience Research in Children's Health, Illness and Disability (ORCHID). Great Ormond Street Hospital NHS Foundation Trust, UK
15. Naomi House & Jacksplace Hospices for Children and Young Adults, UK
16. West of England Academic Health Science Network, Bristol, UK
17. Newcastle University, Newcastle upon Tyne, UK

Contents

1. INTRODUCTION

This BTS Clinical Statement addresses the risk assessment, prevention and management of community-acquired pneumonia (CAP) in people with a learning disability. Each section is summarised with key clinical practice points. The recommendations made are based on a comprehensive review of the published evidence, where available and pertinent, but are predominantly based on expert opinion aimed at providing useful, pragmatic guidance.

Learning disability is the preferred term used in the UK to refer to individuals who have "significantly reduced ability to understand new or complex information, to learn new skills" and a "reduced ability to cope independently which starts before adulthood with lasting effects on development"[1]. Individuals with learning disability represent a widely heterogeneous group of people and can be associated with a broad range of primary diagnoses and comorbidities [2].

There is no definitive record of the number of people with learning disability in England. However, Public Health England (PHE) estimates that in 2015 the population was 1,087,100, including 930,400 adults (approximating 2% of the adult population). The estimated prevalence of learning disability in children and young people (CYP) is 2.5% [3]. Far fewer individuals with learning disability were recorded in health and welfare systems. For example, only 324,291 children and adults were identified as having learning disability on General Practitioner (GP) practice-based registers in 2021 [4]. It is likely that those registered have more severe learning disability or more commonly associated conditions e.g. Down syndrome. This is important, as it means that many individuals with learning disability do not have their diagnosis recognised within health and welfare systems potentially increasing their vulnerability through lack of access to regular health reviews and other relevant public health programmes e.g. vaccination programmes. A series of measures have been introduced by the NHS to help improve identification of people with learning disability [5].

The life expectancy of people with learning disability is reduced compared to the general population; only 37% of adults with learning disability live beyond 65 years of age compared to 85% of the general population [2].

Pneumonia is the commonest cause of death amongst adults with learning disability and is also a common cause of death in CYP with learning disability, with bacterial pneumonia accounting for a significant proportion of these (Adults 24%, Children 21%)[6]. A further 17% of adult and 3% of paediatric deaths in the learning disability population are caused by aspiration pneumonia (AP) [6].

CAP is a major contributor to the increased hospitalisation risk that has been described for people with learning disability and results in longer hospital stays than the general population [7]. People with learning disability also experience increased rates of repeated admission secondary to CAP [8].

As CAP is a major cause of death in people with learning disability, prevention, early detection and pro-active management are key to reducing mortality from avoidable causes [9]. It is important that public sector organisations make reasonable adjustments in their approach or provision to ensure that people with learning disability have equitable access to good quality healthcare [10,11].

SCOPE

Learning disability is variously defined. For the purposes of this statement, learning disability is defined in accordance with the UK Department of Health and Social Care definition[1]. Learning disability is a term that is widely recognised by health and social care professionals in the UK although it is acknowledged that "intellectual disability" is becoming the more widely accepted term internationally [12].

The commonest classification of pneumonia is based on where the patient was when the pneumonia began. Community-acquired pneumonia is the term used to describe an acute infection of the lungs that develops outside the hospital setting in a patient who has not been recently hospitalised. CAP is a heterogeneous disease with multiple potential pathogens including many bacteria and viruses. Microorganisms causing CAP reach the lungs either by inhalation of droplets created by sneezing or coughing from an infected contact or by microaspiration after colonisation of the nasopharynx with a potential pathogen. CAP can also occur as a result of large-volume aspiration (macroaspiration) of colonised oropharyngeal or upper gastrointestinal contents. CAP occurs when there is a defect in normal host defence, a virulent pathogen overwhelms the immune response or there is exposure to a large infectious microbial inoculum. This statement considers all potential CAP aetiologies but does not address CAP secondary to COVID-19 which is covered in detail elsewhere [13].

Although learning disability may be an isolated finding, it is commonly associated with other medical conditions or comorbidities [14]. Learning disability is commonly identified in individuals with neurodisability [15]. Neurodisability describes "a group of congenital or acquired long-term conditions that are attributed to impairment of the brain and/or neuromuscular system and create functional limitations. Conditions may vary over time, occur alone or in combination and include a broad range of severity and complexity. The impact may include difficulties with movement, cognition, hearing and vision, communication, emotion, and behaviour" [16]. Comprehensive reviews and guidelines for the respiratory management of many specific neurodisability subgroups have already been published [17–20]. Whilst this statement does not attempt to replicate their detail, it includes information relevant to this patient group where deemed relevant to CAP.

Annual health checks (AHCs) were introduced for all people 14 years and older with learning disability in an effort to minimise the health inequalities faced by this group [21]. The statement group felt that AHCs provide an important opportunity to integrate a focussed assessment on risk factors for CAP and respiratory morbidity. Consequently, the statement is structured with an emphasis on risk factor identification and instigation of preventive measures.

This statement applies to people with learning disability across all age ranges. CYP refers to any person under the age of 18.

2. Clinical Practice Points

General

- Healthcare professionals should be able to recognise risk factors for CAP in people with learning disability.
- The full assessment for risk factors and the introduction of appropriate preventive measures requires a multidisciplinary approach involving physicians, nurses, physiologists, physiotherapists, speech and language therapists, oral hygienists, dentists, dietitians, radiologists and community learning disability teams.

Engagement and Assessment

- Engaging effectively with the person with learning disability and their parents/carers is fundamental to the early identification of CAP risk factors and the judicious implementation of preventive care and treatment.
- Every encounter is an opportunity for healthcare professionals to build a strong foundation to get it right for all future healthcare.
- Clinical encounters should be well planned to enable reasonable adjustments to be implemented including adequate clinic space, collaboration between health care professionals to minimise visits, additional appointment time and appropriate communication aids.
- A detailed history and clinical assessment of respiratory health should be part of every consultation for people with learning disability (notably at each annual health check) and should be directed at identifying risk factors for CAP and the need for further investigation.

Identifying Risk Factors and Preventive Measures

Lung Function Testing

- Spirometry should be measured as part of the respiratory assessment in all people with learning disability who can perform the manoeuvre.
- Slow Vital Capacity and Peak Cough Flow (PCF) are measures of respiratory muscle strength and should be measured as part of the respiratory assessment in all people with learning disability who have a neuromuscular disorder (NMD) or where there are clinical concerns regarding weak cough or poor secretion clearance.
- Patients with NMD who have a vital capacity of <60% predicted should have an overnight assessment of breathing.
- People with learning disability over 12 years of age who have a PCF <270 L/min should be referred to a specialist respiratory physiotherapist for cough assessment.

Long-Term Assisted Ventilation

- As part of the decision to commence long-term assisted ventilation, people with learning disability and their parents/carers should be fully informed regarding the rationale for assisted ventilation, what it is likely to achieve, its limits and associated potential harms and burdens.
- CPAP or NIV may reduce the rate of respiratory exacerbations in patients with poor secretion management and frequent episodes of CAP.

Airway Clearance

- People with learning disability who have repeated episodes of CAP, a reduced peak cough flow or where there are concerns regarding secretion clearance should be referred to a specialist respiratory physiotherapist for cough assessment and, where indicated, introduction of tailored airway clearance techniques.
- Mechanical insufflation-exsufflation (MI-E) is the treatment of choice in patients with NMD to enhance cough efficacy.
- People with learning disability who cannot clear secretions with conventional therapies (breathing exercises, manual techniques and positioning), should be considered for Positive Expiratory Pressure (PEP) devices, MI-E, high frequency chest wall oscillation or intrapulmonary percussive ventilation.

Physical Activity

- People with learning disability should be encouraged to be as physically active as possible.

Consideration of Prophylactic Antibiotics

- In patients with frequent episodes of CAP, enteral prophylactic antibiotics may be considered following optimisation of modifiable risk factors for CAP and appraisal of individual circumstances including frequency of infection, degree of neurological impairment and the patient's/carers' informed views.
- Azithromycin is the preferred first line enteral prophylactic agent in the absence of contraindications.
- In patients with frequent episodes of CAP and ongoing respiratory tract colonisation with bacteria (notably *Pseudomonas aeruginosa*) a trial of long-term nebulised antibiotics may be considered. A challenge test should be performed prior to initiation.

Aspiration

Eating, Drinking and Swallowing Difficulties

- Eating, drinking and swallowing difficulties (EDS) difficulties may be a contributing factor to CAP.
- Early involvement of speech and language specialists is essential for all those with a potential history of EDS difficulties.

Gastro-oesophageal Reflux Disease (GORD)

- GORD can be difficult to recognise in people with learning disability and may present with atypical symptoms such as rumination and haematemesis
- Where GORD is deemed a potential cause of recurrent AP, referral to a specialist service should be considered for further investigation and management.
- Fundoplication or jejunal feeding may be considered in carefully selected patients with severe GORD and recurrent episodes of aspiration pneumonia where other risk factors have been excluded or optimally treated

Excessive Oral Secretions

- The identification of posterior drooling as a risk factor for CAP can be challenging, necessitating careful history and examination. Frequent coughing, gagging, choking, "wet"/gurgling sounds or daytime anterior drooling that ceases at night may be indicative.
- Optimisation of co-morbidities and other factors contributing to sialorrhoea may help improve saliva control. These include inadequately controlled GORD, nasal obstruction leading to open-mouthed posture, inadequate postural management, poor dental hygiene and medication side effects.
- First line pharmacological management comprises antimuscarinic drugs (hyoscine hydrobromide, glycopyrronium). Dose titration is necessary to avoid drying secretions excessively since lower airway secretions may become thicker and secretion clearance may become difficult.
- If first line pharmacological management is unsuccessful, salivary gland Botulinum A Toxin injection and surgical interventions should be considered in problematic sialorrhoea

Poorly Controlled Seizures

- The diagnosis and management of epilepsy in people with learning disability should be optimised according to national guidance

Oral Health

- Oral health should be regularly assessed in people with learning disability and an oral health care plan developed, ideally in consultation with the person's dentist.
- Good proactive oral care is essential and is most conveniently achieved by brushing teeth and gums with a soft toothbrush at least twice a day with a fluoride toothpaste using non-foaming toothpaste in those with swallowing difficulties to reduce aspiration risk.
- People with learning disability may be reliant on others to provide oral care. This must not be jeopardised if care settings change e.g. on admission to hospital.

Nutritional Considerations

- An appraisal of nutritional status, including measurement of height and weight, should be a fundamental part of the assessment of people with learning disability.

- The diagnosis, investigation and management of nutritional disorders in people with learning disability should be optimised according to national guidance.

Vaccination

- Vaccine history should be reviewed at every opportunity.
- Annual influenza vaccination is recommended for all people with learning disability.
- Pneumococcal vaccination should be considered as specified by the Joint Committee on Vaccination and Immunisation recommendations.
- People with learning disability should be considered as high priority in all vaccination programmes for seasonal respiratory infections.

Smoking

- Every opportunity should be taken to review smoking and environmental tobacco smoke (ETS) exposure status.
- Smoking cessation advice and referral onto smoking cessation services should be offered to all people with learning disability who are current smokers and, where significant ETS exposure is identified, to their parents/carers.

Aetiology, Diagnosis, Investigation & Management of Community Acquired Pneumonia

Early Recognition

- Early recognition of CAP can be challenging in people with learning disability. Soft signs, such as changes to baseline alertness and mobility, should be viewed with a high index of suspicion.

Diagnosis

- A comprehensive history should be sought from the patient, using communication aids when required, with additional information gathering from parents and carers.
- The severity of CAP should be assessed to guide treatment and decision making around requirement for hospital assessment and admission. In adults with learning disability, the CRB-65 or CURB-65 should be used in conjunction with clinical judgement. In CYP, clinical features such as respiratory rate, difficulty in breathing and hydration status can be used to assess severity.

Investigation

- A chest radiograph should be performed in all adults requiring hospital assessment with suspected CAP and should be considered in CYP with features of severe or complicated pneumonia.
- In CYP with severe CAP and adults with moderate to severe CAP (for example CURB 65>2), blood testing and microbiological investigations including sputum culture and sensitivity, blood cultures and pneumococcal urine antigen should be undertaken to help guide treatment.
- In all CYP and adults being assessed for suspected CAP, pulse oximetry should be undertaken as it can be a useful adjunct to clinical decisions regarding site of care and need for further investigation.
- For hospitalised adults with CAP, NEWS2 (or an equivalent early warning score) should be calculated and tracked throughout hospital assessment and admission to monitor for signs of deterioration and to facilitate timely clinical response.

Management

- Antibiotic therapy should be guided by clinical severity, known pathogens and allergies.
- In CYP with CAP, supplementary oxygen should be administered to maintain oxygen saturations 93-98%.
- In adults, target oxygen saturations between 94-98% should be used in people where there is no risk of hypercapnic respiratory failure although more conservative oxygen targets can also be considered, in keeping with local guidance. For those with risk factors for hypercapnic respiratory failure (such as scoliosis, neuromuscular disease, chronic obstructive pulmonary disease or obesity), oxygen saturations should be maintained between 88-92%.
- In patients with retention of secretions, early involvement of chest physiotherapy should be considered.

Consideration of Risk Factors and Follow Up

- An episode of CAP should trigger a full respiratory assessment including detailed clinical history and examination, review of risk factors and consideration for pertinent investigations.
- Follow up should be arranged for all patients who present with CAP and have been identified as having risk factors so that a holistic assessment of the patient's needs can be made, and risk factors addressed.

Palliative Care Considerations

- All health professionals should be able to provide quality palliative care with support from specialist palliative care teams where necessary.
- Palliative care can be delivered alongside active treatment in a parallel planning approach. The two are not mutually exclusive.
- Palliative care focusses on quality of life. Developing a personalised plan in advance can help to record a patient's usual quality of life, priorities and goals. The patient should be supported to contribute to this as much as they are able along with family or friends who can advocate for them.

3. Glossary of Terms

ACT	Airway Clearance Technique
AHC	Annual Health Check
AP	Aspiration Pneumonia
BMI	Body Mass Index
BTS	British Thoracic Society
CAP	Community-Acquired Pneumonia
CF	Cystic Fibrosis
CP	Cerebral Palsy
CPAP	Continuous Positive Airways Pressure
CYP	Children and Young People
DMD	Duchenne Muscular Dystrophy
DS	Down Syndrome
EDS	Eating, Drinking and Swallowing
ENT	Ear, Nose and Throat
ETS	Environmental Tobacco Smoke
FEES	Fibreoptic Endoscopic Evaluation of Swallow
GORD	Gastro-Oesophageal Reflux Disease
GP	General Practitioner
HFNC	High Flow Nasal Cannula
JCVI	Joint Committee on Vaccination and Immunisation
LPA	Lasting Power of Attorney
LRTI	Lower Respiratory Tract Infection
MDT	Multi-Disciplinary Team
MECC	Making Every Contact Count
M I-E	Mechanical Insufflation-Exsufflation
MIP	Maximum Inspiratory Pressure
MRSA	Methicillin-Resistant *Staphylococcus aureus*
NIV	Non-Invasive Ventilation
NM	Neuromuscular
NMD	Neuromuscular Disorder
OSA	Obstructive Sleep Apnoea
PA	*Pseudomonas Aeruginosa*
PCF	Peak Cough Flow
PCR	Polymerase Chain Reaction
PPI	Proton Pump Inhibitor
PWS	Prader-Willi Syndrome
SNIP	Sniff Nasal Inspiratory Pressure
SLT	Speech and Language Therapist
SMA	Spinal Muscular Atrophy
VFS	Videofluoroscopy of swallow
WHO	World Health Organization

4. Risk Factors

Important risk factors have been identified that increase the likelihood of an individual with learning disability developing CAP. These are summarised in table 1. It is important to consider these risk factors whenever a person with learning disability is assessed.

Risk Factor (*- modifiable)	Comments
Sleep Disordered Breathing*	Although not specifically studied in people with learning disability, non-interventional retrospective analyses support an association between obstructive sleep apnoea (OSA) and CAP [22,23]. Several retrospective cohort studies have demonstrated an increased rate of CAP in patients with NMD and nocturnal hypoventilation with a reduction following appropriate treatment [24,25].
Past history of respiratory infections	Previous episodes of CAP indicate an increased risk of subsequent CAP [26]. This risk is of particular note in Cerebral Palsy (CP) where a respiratory hospital admission is associated with a >50% likelihood of a further hospitalisation for CAP in the subsequent 12 months[27,28].
Reduced Mobility/ Impaired Motor Function*	Reduced mobility and impaired motor function have been identified as a risk factor for pneumonia in many disorders associated with learning disability including CP [27,29]. Physical activity and mobility reduce more rapidly in older adults with learning disability compared to the general population [30,31].
Eating, Drinking & Swallowing Difficulties	People with learning disability are at increased risk of swallowing difficulties. Population studies have found 8.1% to 11.5% of adults known to formal learning disability services present with swallowing difficulties[32,33] although this is likely an underestimation due to diagnostic issues and selection bias [34]. Eating, Drinking and Swallowing difficulties are a recognised risk factor for CAP in people with learning disability [35].
Gastro-Oesophageal Reflux Disease (GORD)*	GORD has been identified as a risk factor for CAP [36], most notably in those with neurodisability [27]. Clinical features that may indicate GORD include feeding refusal, failure to thrive, heartburn/chest pain and epigastric pain.
Sialorrhoea*	Sialorrhoea is increased saliva in the mouth, which can be either anterior, resulting in drooling, or posterior, where the saliva pools in the oropharynx and posterior pharynx. This can result in aspiration and aspiration pneumonia [37].
Poorly controlled epilepsy*	Seizures are associated with an increased risk of aspiration, including of vomit or saliva[38]. Medications used to control or terminate seizures can lead to reduced muscle tone or drowsiness, further increasing the risk of aspiration [18].
Bacterial colonisation*	Bacterial colonisation, including with Gram-negative organisms such as *Pseudomonas aeruginosa*, is associated with more severe CAP, including increased rates of hospitalisation and critical care admission and prolonged hospitalisations [39,40].
Immunodeficiency	Immunodeficiency is a feature of several learning disability-associated syndromes such as Down and Di George. Deficiencies in both humoral and cellular immunity have been identified in Down syndrome, for example, and are felt to at least partially explain the increased risk of CAP that has been described in these patients [41].
Tracheostomy	Infection and colonisation are common long-term complications of tracheostomies in both children and adults, with *Pseudomonas aeruginosa* (PA) colonisation in up to 90% of children with tracheostomies [42]. Presence of a tracheostomy has been identified as an independent risk factor for developing PA associated CAP [40].

Oral Health*	People with learning disability experience more problems with their oral health than the general population [43]. Poor oral care and decaying teeth are risk factors for CAP in the elderly with some evidence in younger age groups [44,45].
Nutritional Status*	People with learning disability have a higher incidence of both obesity and being underweight when compared with the general population [46–48]. Being underweight is associated with an increased risk of CAP [49]. Obesity may lead to reduced mobility and secondary problems such as obstructive sleep apnoea and gastro-oesophageal reflux disease which in turn may impact chest health [50].
Smoking / Environmental Tobacco Smoke (ETS) exposure*	Smoking is associated with an increased risk of developing CAP [51]. ETS exposure in childhood is associated with an increased risk of hospitalisation for CAP and increased severity of disease once hospitalised [52,53].
Presence of Comorbid Conditions	Comorbid conditions including chronic heart disease, chronic obstructive pulmonary disease (COPD), diabetes mellitus, cerebrovascular diseases, dementia, chronic liver disease and chronic renal disease are recognised risk factors for CAP [54].

Table 1 Risk factors for CAP in people with learning disability

5. Engagement and Assessment

Engagement

Engaging effectively with people with learning disability and their families is key to early identification of CAP risk factors, implementation of proactive management to mitigate against CAP, and the delivery of high-quality care and treatment that is tailored to their needs. A recent model for the delivery of equitable care to children and young people with learning disability provides a good framework for considering how best to engage with patients with learning disability [55]. Central to this is for staff and teams to work in partnership with patients, parents, family members, carers and community partners from the outset to ensure that appropriate reasonable adjustments are made in the context of learning disability to enable comprehensive accurate clinical assessment and appropriate care and treatment [56]. This may include accounting for learning disability in family carers. The principle of making every contact count (MECC) [57] is vital.

Effective communication is key to good engagement. This includes understanding how the person usually communicates and providing information in accessible formats, for example using easy read materials. Mencap provide helpful guidance on communicating with people with learning disability [58]. Used effectively, hospital passports or "All About Me" documents are useful to enable people with learning disability and families/carers to share information with health staff about their needs and help them make the necessary reasonable adjustments to the care and treatment they provide [59].

It is important to recognise the legal duty of healthcare services to consider the needs of all people with disabilities in the way they organise their buildings, policies and services as set out in The Equality Act 2010 [60]. Reasonable steps need to be made to avoid disadvantage arising from any provision, criterion, practice, physical feature, or lack of auxiliary aids.

Assessment

Every opportunity should be taken to assess individuals with learning disability for risk factors for CAP (table 1 and Section 3). A thorough history and clinical examination are the cornerstone of assessment. Figure 1 outlines important elements of the assessment and highlights relevant findings which may suggest the need for further investigation.

Careful forward planning is important to ensure appointments are effectively utilised. Appropriate reasonable adjustments should be made such as easy read documents, keeping language simple and providing longer appointments if possible. The potential for support from their carer, family member or the community learning disability nursing service should be considered.

History and review of systems

- Past respiratory history including episodes of previous pneumonia, requirement for antibiotics, past hospital admissions, prescribed respiratory physiotherapy and use of assisted ventilation
- Medication review including anti-epileptic drugs, psychotropic medication and regular antibiotics
- Review of long-term conditions such as epilepsy, diabetes or renal disease
- Specific questions regarding eating, drinking and swallowing to include choking, coughing when eating, or food 'getting stuck'
- Specific review of symptoms which may identify risk factors for aspiration such as drooling, symptoms of gastro-oesophageal reflux (heartburn, waterbrash, vomiting, rumination etc) or uncontrolled seizures
- History suggestive of sleep disorder breathing such as excessive daytime sleepiness, habitual loud snoring, witnessed apnoea during sleep, nocturia, morning headache or nausea
- History suggestive of impaired airway clearance including history of difficulty clearing secretions and crackles, bubbling sounds or coarse wheezing at the mouth during coughing or deep breathing
- Dental history (including oral hygiene regimen and history of previous dental care)
- Immunisation history (specifically influenza and pneumococcal vaccines)
- Physical activity (any recent changes)
- Smoking history including exposure to smoke from family members and carers

Examination

Examination should consider:

- Weight, height and BMI calculation. Consider the use of wheelchair scales.
- Baseline observations such as pulse rate, temperature, oxygen saturations and respiratory rate to facilitate identification of physiological change in acute illness
- General assessment of posture (including scoliosis) and muscle tone
- Assessment for cough strength and character
- Assessment for excessive oral secretions (drooling, gurgling)
- Assessment of oral health including teeth (decay, acid erosion), gums, tongue and buccal mucosa
- Presence of tracheostomy, enteral feeding tubes
- Signs of sputum retention including crackles or bubbling sounds at the mouth during coughing or deep breathing, palpable secretions, audible crackles or wheeze on chest auscultation (which may clear after coughing)

Figure 1 Clinical assessment

Assessment in Primary Care

The primary care team have an important role to play to identify those at increased risk of community acquired pneumonia and offer appropriate interventions and referrals as needed.

Figure 2 highlights key considerations for primary care clinicians when assessing people with learning disability.

- It is estimated that 3 in every 4 patients with learning disability are not identified on the learning disability register – vigilance is important to ensure patients are appropriately registered. Consider using an inclusion tool [61] and national guidance [5] to help identify those who should be registered.
- Life expectancy is significantly reduced compared to the general population and many die of preventable causes: over 40% of adults with learning disability die of pneumonia
- Those who have had previous CAP are at increased risk of further episodes
- Implement annual health checks for all people with learning disability; ensure reasonable adjustments are considered to facilitate the consultation such as longer appointments
- Use annual health checks as an opportunity to offer health promotion such as pneumococcal or influenza vaccination.
- Make every contact count - review long term conditions and identify risk factors for CAP that are amenable to intervention:
    - poor cough efficacy
    - sleep-disordered breathing
    - eating, drinking and swallowing difficulties
    - gastro-oesophageal reflux
    - excess saliva
    - poor seizure control
    - dental hygiene
    - reduced physical activity
    - underweight or obesity
    - smoking and environmental tobacco smoke exposure
- Consider referral for further assessment e.g. speech and language therapists for a swallowing assessment
- Summarise findings and recommendations in a health action plan

Figure 2 Key considerations for primary care clinicians

Annual Health Checks

An annual review by the general practitioner, the annual health check (AHC), was introduced across the UK for all people with learning disability aged over 14 years to address health inequalities faced by people with learning disability. AHCs are associated with a reduction in emergency admissions for conditions such as pneumonia [62]. LeDeR data indicate that there is a 1.5 times greater likelihood of a person with a learning disability dying if they have not had an annual health check in the year prior to their death [63].

AHCs are a systematic review during which new health problems can be identified, management of long-term conditions optimised, and preventative interventions offered [64,65]. The value of the AHC is greatly enhanced by appropriate preparation for the appointment [65] and to improve uptake it is important to consider reasonable adjustments such as longer appointments [56,64,66,67]. An easy read pre-check questionnaire sent to the patient or the family, or carer if relevant, can enable the collection of information such as vaccination history and recent dental review findings and can

identify priority areas for review such as eating, drinking and swallowing difficulties [68,69]. Electronic templates can be used to support the collection of information [64,66].

A full assessment should be completed (see figure 1). AHCs also provide an opportunity for health promotion such as advice on healthy eating or exercise.

At the end of the AHC, a health action plan should be generated [68] and held by the person with learning disability to summarise what has been discussed including recommendations (e.g. adjustments to medications) and details regarding any referrals. This also provides an opportunity to share health promotion information. It is essential that this information is shared in a format accessible to each individual e.g. easy read formats [66].

Clinical Practice Points

- Engaging effectively with the person with learning disability and their parents/carers is fundamental to the early identification of CAP risk factors and the judicious implementation of preventive care and treatment
- Every encounter is an opportunity for healthcare professionals to build a strong foundation to get it right for all future healthcare
- Clinical encounters should be well planned to enable reasonable adjustments to be implemented including adequate clinic space, collaboration between health care professionals to minimise visits, additional appointment time and appropriate communication aids
- A detailed history and clinical assessment of respiratory health should be part of every consultation for people with learning disability (notably at each annual health check) and should be directed at identifying risk factors for CAP and the need for further investigation.

6. Identifying Risk Factors and Preventive Measures

Whilst the risk factors for CAP are well described (Table 1), there is often little or only weak evidence for the impact of intervention on subsequent CAP. There is, however, broad consensus regarding the contribution of each risk factor to the pathophysiology of CAP and mitigating strategies that can be employed.

This section provides strategies to (1) enable early recognition of CAP risk factors and (2) implement pragmatic proactive management to address each risk factor.

Lung Function Testing

Spirometry

Spirometry is an important part of the respiratory assessment providing valuable information regarding pulmonary function and helping identify patients that warrant further investigation. A reliable forced expiratory manoeuvre may not be possible due to weakness, poor coordination or insufficient comprehension. In these cases, a slow vital capacity may often be obtained by asking the patient to breathe in as deeply as possible and then expire slowly for as long as possible into the spirometer [70]. Where the patient cannot achieve a seal around a conventional mouthpiece, a flanged mouthpiece or an appropriately sized facemask held tightly over the nose and mouth can be employed [71]. Vital Capacity (VC) can be an effective means of monitoring disease progression and has been shown to strongly correlate with the number of chest infections and days of antibiotic treatment in the preceding year in children with a variety of muscle diseases [72]. VC is also predictive of sleep-disordered breathing and hypoventilation in patients with NMD [73]. Overnight monitoring of breathing is recommended for patients with NMD that have a VC of <60% predicted [19]. VC obtained in the supine position may help to assess diaphragmatic function in patients with NMD [74].

Predictive values rely on an accurate standing height which may be unreliable or unobtainable particularly in non-ambulant patients or those with a significant scoliosis. Arm span or ulna length can be used as surrogate markers of height in this context [19].

Peak Cough Flow (PCF)

Individuals with learning disability may have an impaired cough due to respiratory and bulbar muscle weakness particularly in those with neurodisability. A PCF of <270 L/min is associated with an increased risk of complications from common respiratory infections in adults with NMD [25]. This cut-off value is probably also relevant to children > 12 years of age although, for younger children, similar risk thresholds have not been established [19]. PCF is recommended as part of the assessment of secretion clearance in children over the age of 12 years with NM weakness [19] or where there are clinical concerns regarding cough efficacy. Where PCF cannot be performed but the patient can perform a FVC, formal cough evaluation should be considered for patients with a FVC of <75% predicted as this can potentially indicate the need for interventions to improve cough strength [75].

Tests of Respiratory Muscle Strength

Assessment of respiratory muscle strength, including sniff nasal inspiratory pressures (SNIP) and maximum inspiratory pressures (MIP), can provide further valuable data to guide management [76,77].

Both SNIP and MIP correlate strongly with VC [78–80] and may be helpful in certain patients. For example, patients with bulbar dysfunction may have difficulty creating an adequate seal with a mouthpiece for spirometry but may be able to perform a SNIP manoeuvre[81].

Clinical Practice Points

- Spirometry should be measured as part of the respiratory assessment in all people with learning disability who can perform the manoeuvre.
- Slow Vital Capacity and Peak Cough Flow (PCF) are measures of respiratory muscle strength and should be measured as part of the respiratory assessment in all people with learning disability who have NMD or where there are clinical concerns regarding weak cough or poor secretion clearance
- Patients with NMD who have a vital capacity of <60% predicted should have an overnight assessment of breathing
- People with learning disability over 12 years of age who have a PCF <270 L/min should be referred to a specialist respiratory physiotherapist for cough assessment

Overnight Monitoring of Breathing

The overall prevalence of sleep disorders is elevated in people with learning disability [82] particularly in those with an associated neurodisability [83–85]. Overnight sleep monitoring enables early recognition of hypoventilation and upper airway obstruction.

The relationship between obstructive sleep apnoea (OSA) and CAP has not been specifically studied in people with learning disability, but an association is increasingly recognised in other groups [22,23]. Similarly, the relationship between hypoventilation and the reduced incidence of CAP following treatment has only been studied in patients with NMD [24,25]. The statement group felt that the early recognition of both OSA and hypoventilation was important despite a lack of strong evidence in relation to CAP prevention particularly given the other potential benefits of appropriate treatment.

Indications for overnight sleep monitoring are listed in Table 2.

Indication	Notes
Individuals with NMD with a vital capacity <60%	The incidence of nocturnal hypoventilation has been found to increase with reduced vital capacity in a variety of neuromuscular diseases [78,86–88]
Symptoms of OSA	Excessive daytime sleepiness, habitual loud snoring, witnessed apnoea or gasping or choking during sleep, nocturia
Symptoms of hypoventilation	Excessive daytime sleepiness, morning headache, morning nausea, poor concentration, reduced performance in school, changes in behaviour, reduced appetite.
Individuals with NMD who lose the ability to walk due to progressive weakness or children who never attain the ability to walk	Inability to walk is a cardinal sign of moderate to severe muscle weakness [19]
Down syndrome (DS)	An estimated 30-79% of children with DS have OSA with an even higher prevalence reported in adults [89,90]. Overnight sleep monitoring is recommended, using at least oximetry, at least once in infancy then annually [89].
Prader-Willi Syndrome (PWS)	Sleep disordered breathing is a common finding in PWS [90]. All children with PWS should be screened with oximetry and capnography on an annual basis [89].

Table 2 Indications for overnight monitoring of breathing in people with learning disability

Several techniques are available to assess sleep-disordered breathing with varying complexity, degree of patient inconvenience and cost. These include pulse oximetry, oxycapnography, respiratory polygraphy and polysomnography. The choice of which technique is the most appropriate depends on multiple factors including patient compliance and is discussed in detail elsewhere [19,91,92].

Long-Term Assisted Ventilation

Assisted ventilation has a useful role in several clinical scenarios. Its principal uses are for the correction of OSA and the treatment of hypoventilation and may also be used as a long-term intervention to prevent atelectasis and segmental collapse where secretion management is poor and respiratory exacerbations are frequent.

Non-invasive ventilation (NIV) refers to non-invasive positive pressure ventilation with two levels of pressure applied during the respiratory cycle and continuous positive airways pressure (CPAP) refers to the non-invasive application of positive airway pressure at a continuous level [107].

The benefits of long-term assisted ventilation are well established in several conditions associated with learning disability, most notably those with NMD. NIV and CPAP can be delivered by several different interfaces including face mask, nasal mask, nasal pillows or mouthpiece. Patients with learning disability may initially struggle to tolerate NIV and CPAP particularly as they may not be able to understand the rationale for treatment despite careful explanation. They may develop a

conditioned anxiety associating the sight, sound and sensation with mask discomfort and physiologic arousal [108]. Non-invasive interfaces delivering both CPAP and NIV can insufflate the stomach and may aggravate an existing vulnerability to reflux aspiration. Improved interface design has made it possible for NIV and CPAP to be well tolerated by most patients. Motivational and behavioural interventions may help improve adherence [108,109].

It should be explained what the intervention is likely to achieve, but also its limits, and any further escalations that may be indicated. It is also important to outline the potential risks and burdens of the intervention including any increased limitations on life at home or reduction in choice about community settings that can support certain technologies. This is an opportunity to record the patient's priorities and preferences about this and future interventions, and to explore any circumstances under which they may wish to have the intervention withdrawn.

CPAP

Where OSA is identified, there should be an assessment of any potential causative factors (eg adenotonsillar hypertrophy, upper airway hypotonia, craniofacial morphology, obesity) and careful appraisal of all treatment options. In CYP, a referral to the ear, nose and throat (ENT) specialist team should be considered for upper airway evaluation and consideration for surgical management (eg adenotonsillectomy). A nasopharyngeal airway may be indicated particularly in those unable to tolerate CPAP or with craniofacial abnormalities.

A trial of CPAP is indicated where surgical management is deemed inappropriate (due to patient age, anatomical findings, surgical risk or patient preference) or where OSA persists despite surgery. CPAP maintains patency of the upper airway by overcoming the critical closing pressure of the pharynx [110]. Pressure titration is important to optimise CPAP efficacy and minimise unnecessary pressure effects. Regular follow up is important once CPAP has been established to monitor adherence and efficacy and to optimise equipment/pressure level as necessary.

NIV

NIV is indicated where there is evidence of hypoventilation. In patients with NMD, night-time NIV has been demonstrated to correct nocturnal hypoventilation and reverse daytime hypercapnia [86,111,112] with some evidence for a reduction in frequency of hospital admission for CAP [111,113,114] and an increased life expectancy in Duchenne Muscular Dystrophy and Spinal Muscular Atrophy [115,116].

A wide array of ventilators are available for home ventilation providing multiple different modes of ventilation. The principal aim is to match the ventilator/ventilation mode to each patient to optimise improvements in respiratory pathophysiology whilst ensuring maximal patient comfort and patient-ventilator synchrony. Clinically stable patients can be effectively and safely commenced on NIV in the home setting [117]. Once NIV has been established, overnight sleep monitoring should be performed to ensure hypoventilation has been effectively alleviated. The frequency of subsequent review and sleep monitoring will depend on individual circumstances including age, clinical stability and compliance with NIV. Review should include assessment of NIV efficacy and adherence, evaluation of need for equipment/interface modifications and monitoring for potential adverse effects such as skin breakdown and midface hypoplasia.

Some patients (particularly those with severe weakness) may require daytime NIV due to hypercapnia during wakefulness that does not respond to nocturnal NIV alone. This can be delivered via the same interfaces that are used at night-time or via a mouthpiece. Mouthpiece ventilation enables the patient to define their own ventilatory pattern [118].

Tracheostomy / Invasive Ventilation

Insertion of a tracheostomy may be considered for ongoing care in certain circumstances:

1. Requirement for prolonged daytime ventilation
2. Failure to extubate following a period of invasive ventilation due to an acute deterioration
3. Severe bulbar dysfunction resulting in episodes of recurrent aspiration (to facilitate management of secretions)
4. NIV fails to correct hypoventilation and/or hypoxia

The decision to proceed to a tracheostomy must be fully informed with clear communication regarding the benefits, but also the burdens. For some having a tracheostomy can be transformative making eating easier, improving social interaction (without the requirement for obstructive facial interfaces) and providing stability in respiratory health. However, for others the burden of constant intervention (with suction etc), loss of ability to communicate spontaneously verbally, and dependence on trained carers (often with a prolonged stay after insertion whilst a care package is established) means it can be repressive. Potential tracheostomy complications should also be considered including granuloma and fistula formation, increased secretions, swallowing difficulties and increased respiratory infections [119]. Advance care planning can play a key role in part of this decision-making process in encouraging patients and their families to consider and articulate their wishes.

Clinical Practice Points

- As part of the decision to commence long-term assisted ventilation, people with learning disability and their parents/carers should be fully informed regarding the rationale for assisted ventilation, what it is likely to achieve, its limits and associated potential harms and burdens.
- CPAP or NIV may reduce the rate of respiratory exacerbations in patients with poor secretion management and frequent episodes of CAP.

Airway Clearance

Secretions and cough

Ineffective clearance can result in retained secretions which may cause airways obstruction, atelectasis and segmental collapse and may become infected.

Mucus is transported under normal circumstances from the peripheral airways into the pharynx by cephalad-bias airflow and the mucociliary escalator mechanism. Secretion movement can be enhanced using the following three strategies: increasing lung volumes, increasing expiratory flows and utilisation of collateral ventilation. These are the main principles of airway clearance techniques (ACTs). Once secretions reach the proximal airways they can be cleared with an effective cough [93]. An effective cough comprises 3 components: deep inspiration, rapid closure of the glottis with contraction of abdominal and intercostal (expiratory) muscles [94] and explosive decompression upon glottic opening [95]. If one or more of these components are impaired the cough will be less effective [96].

673

674 *Assessment and Airway Clearance Techniques*

675 Where there is suspicion of an ineffective cough, a full assessment should be performed [18]. This
676 involves assessment of whether: (a) there is a deep breath in without paradoxical movement and (b)
677 there is a good expiratory muscle contraction resulting in an audible rapid expulsion of air [97]. Bulbar
678 function can be assessed by asking the patient to repeat e, e, e, e, to determine whether the vocal
679 cords are opening and closing effectively. Cough strength can be measured using PCF (see above)[75].

680 People with learning disability, particularly those with associated neurodisability, may have impaired
681 airway clearance for several reasons. All 3 components of cough can be affected. The inspiratory
682 phase can be impaired due to diffuse muscle weakness and alterations of the mechanical properties
683 of the lungs and chest wall [98]. Adequate intrapleural pressures for the rapid expulsion phase may not
684 be possible due to weak accessory muscles of expiration. Glottic closure can be impaired due to
685 bulbar impairment and prevented in patients with a tracheostomy.

686 Appropriate medical management of secretions should occur prior to instigation of Respiratory
687 Physiotherapy techniques. Adequate hydration is important to assist mucociliary clearance. If
688 secretions remain tenacious despite hydration, nebulised saline can be effective [99]. Excessive
689 secretions may impact the efficacy of ACTs and should also be addressed (see "Excessive Oral
690 Secretions" section below).

691 Respiratory Physiotherapy should be targeted to loosening secretions and moving them to the
692 central airways (peripheral ACTs) or clearing from the central airways and therefore enhancing
693 cough (proximal ACTs) [75,100]. See Appendix 1 for a description of techniques that may be beneficial to
694 individuals with learning disability.

695 People with learning disability may have difficulty understanding the specific manoeuvres required
696 of a proposed technique. The physiotherapist needs to gauge the most effective method for
697 introducing the technique. This may be via demonstration, explanation at their cognitive level or by
698 playing games. In those individuals where distress occurs despite attempts to acclimatise to the
699 technique, choosing a technique that requires no co-operation may be the most effective.

700 As no one treatment technique fits all, Figure 3 highlights treatment options that may be
701 appropriate depending on the severity of learning disability and/or respiratory muscle weakness. A
702 simple technique may often be more appropriate and effective than a complex piece of equipment.
703 Often the physiotherapist will cycle between peripheral ACT and proximal ACT so that secretions are
704 repeatedly moved up into the central airways, coughed and cleared.

705 Provision of a suction machine should be considered where ACT strategies fail or for those who are
706 unable to co-operate with techniques or have a severely weak cough and/or bulbar insufficiency.
707 Depending on the degree of impairment oral and or nasopharyngeal suction should be taught to
708 parents/carers [18].

709 Provision of respiratory equipment for individuals with learning disability often occurs from hospital-
710 based services. It is therefore essential that there is close communication between hospital and
711 community services. Where community services offer rapid respiratory assessment to people with
712 learning disability, the provision for a loan device during times of illness may be useful [101].

713

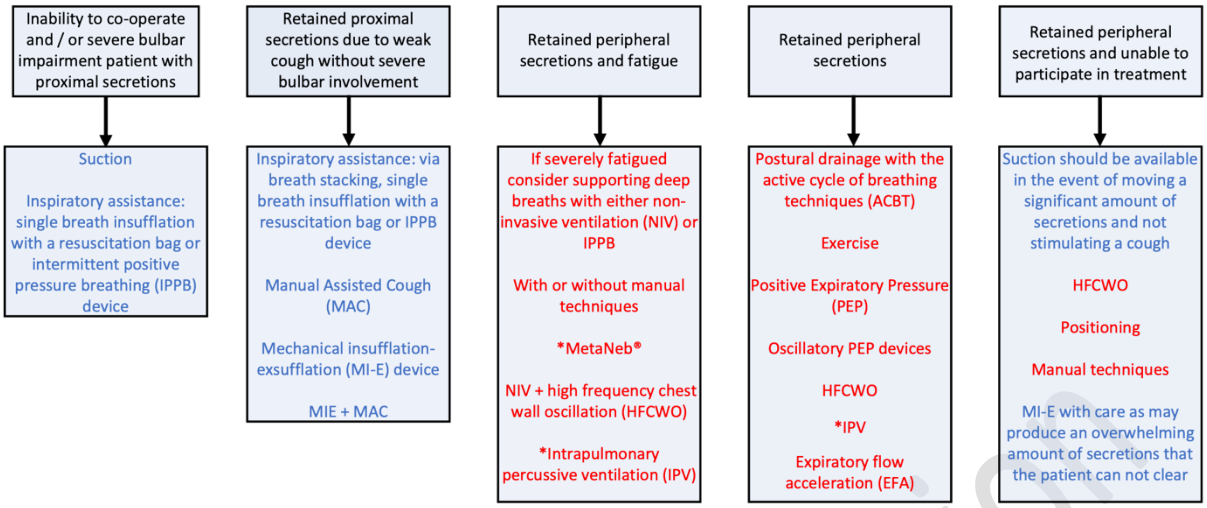

Figure 3.

Respiratory physiotherapy options for patients with learning disability and retained secretions. Techniques in blue are proximal airway clearance techniques and techniques in red are peripheral airway clearance techniques.

Note *intrapulmonary percussive ventilation (if available) and the *MetaNeb® are used in the acute settings

Abbreviations: IPPB= Intermittent Positive Pressure Breathing; MAC= Manual Assisted Cough; MI-E= Mechanical Insufflation-Exsufflation; NIV= Non-Invasive Ventilation; HFCWO= High Frequency Chest Wall Oscillation; ACBT= Active Cycle of Breathing Techniques; PEP= Positive Expiratory Pressure; EFA= Expiratory Flow Acceleration

Clinical Practice Points

- People with learning disability who have repeated episodes of CAP, a reduced peak cough flow or where there are concerns regarding secretion clearance should be referred to a specialist respiratory physiotherapist for cough assessment and, where indicated, introduction of tailored airway clearance techniques.
- Mechanical insufflation-exsufflation (MI-E) is the treatment of choice in patients with NMD to enhance cough efficacy.
- People with learning disability who cannot clear secretions with conventional therapies (breathing exercises, manual techniques and positioning), should be considered for PEP devices, MI-E, high frequency chest wall oscillation or intrapulmonary percussive ventilation.

Physical activity

The health benefits of exercise and its impact on respiratory function are well recognised [102]. Exercise increases respiratory flow and has been shown to promote the clearance of respiratory secretions from the peripheral airways [103].

People with learning disability have significantly lower physical activity levels than the general population [104,105] due to multiple logistic, social and personal barriers [106]. Despite a lack of direct evidence of the impact of exercise on rates of CAP, the statement group agreed that physical activity was an important contributor to respiratory health and should be optimised in people with learning disability.

Community Physiotherapists are best placed to provide exercise programs for individuals with learning disability.

Clinical Practice Points

- People with learning disability should be encouraged to be as physically active as possible

Consideration of prophylactic antibiotics

Currently, there is no direct evidence that prophylactic antibiotics reduce the frequency or severity of CAP in people with learning disability [120] although a large randomised trial in children with neurological impairment is currently ongoing [121]. However, the statement group agreed that enteral prophylactic antibiotics should be considered in patients with frequent pneumonia, following assessment/management of modifiable risk factors for CAP and appraisal of individual circumstances including frequency of infection (particularly >3 episodes of CAP or >2 hospitalisations per year), degree of neurological impairment and the patient's/carers' informed views [17]. Some support for this approach is derived from cystic fibrosis (CF) and non-CF bronchiectasis where the use of long-term macrolides appears to be safe and effective [122,123]. Azithromycin is the preferred first line prophylactic agent benefiting from good tissue penetration, broad antibacterial activity, significant anti-inflammatory properties and a long half-life (thus enabling reduced treatment burden through 3 times a week dosing) [124]. Appropriate safety precautions should be employed prior to the initiation of treatment including ensuring non-tuberculous mycobacteria are not present prior to initiation of treatment and obtaining baseline ECG (to assess QTc interval) and liver function tests in adults [124]. Baseline LRTI rate should be established before commencing prophylaxis with efficacy reviewed 6 monthly and consideration of stopping or switching prophylaxis agent if no benefit is demonstrated. Monitoring of cultures and sensitivity from available respiratory specimens may help direct choice of antibiotic for ongoing prophylaxis.

Chronic pulmonary colonisation including with Gram-negative organisms such as PA should be considered in patients with learning disability who have recurrent LRTIs. PA in chronic airways disease is associated with poorer outcomes [125]. Whilst the true incidence of bacterial colonisation in certain learning disability patient groups is debated [17], it is apparent that short-lived oropharyngeal colonisation with bacteria (including Gram-negatives) regularly occurs at the time of viral upper respiratory tract infection [126]. However, eradication should be pragmatically considered if there is recurrent isolation of a certain bacteria, particularly PA. In patients with frequent pneumonias and ongoing colonisation, a trial of long-term nebulised antibiotics may be considered [127–129]. A lower threshold for this treatment strategy should be employed for those with a tracheostomy who are particularly at risk of colonisation (PA most commonly) [130]. Consideration of potential side effects should be made, including that of bronchospasm and a challenge test is advised before initiation [122].

For patients with recurrent chest infections, or daily sputum production, investigation for underlying bronchiectasis is recommended [122].

Clinical Practice Points

- In patients with frequent episodes of CAP, enteral prophylactic antibiotics may be considered following optimisation of modifiable risk factors for CAP and appraisal of individual circumstances including frequency of infection, degree of neurological impairment and the patient's/carers' informed views.
- Azithromycin is the preferred first line enteral prophylactic agent in the absence of contraindications.
- In patients with frequent episodes of CAP and ongoing respiratory tract colonisation with bacteria (notably Pseudomonas aeruginosa) a trial of long-term nebulised antibiotics may be considered. A challenge test should be performed prior to initiation.

Aspiration Pneumonia

Aspiration of upper airway secretions, refluxate, saliva, food or drink can cause CAP. Aspiration Pneumonia (AP) is covered in detail in the accompanying BTS Aspiration Pneumonia statement [131].

The four principal risk factors for aspiration in learning disability are eating, drinking and swallowing difficulties, gastro-oesophageal reflux, excessive oral secretions and poorly controlled seizures.

Eating, Drinking and Swallowing Difficulties

Eating, drinking and swallowing (EDS) difficulties are common among populations who have neurological, muscular, physiological or structural impairment. Swallowing is a complex integrated series of neurological and physiological events. Problems may occur in the mouth, in the pharynx, larynx, or in the oesophagus [132].

People with learning disability are at increased risk of EDS difficulties although its true prevalence has not been established. Population studies have found 8.1% to 11.5% of adults known to formal learning disability services present with EDS difficulties [32,33] although this is likely an underestimation due to diagnostic issues and selection bias [34].

The highest rates of EDS difficulties have been found in those with motor impairment [133], increased severity of learning disability [134,135] and CP. Approximately 60% of people with CP have EDS difficulties [136,137].

EDS difficulties are associated with significant health risks for people with learning disability including pneumonia, chronic lung disease, asphyxia, obstructive sleep apnoea and hypoxemia during oral feeding [136,138–141]. EDS difficulties also have a significant psychosocial impact with patients feeling ashamed, humiliated and isolated [142].

In 2004, the NHS National Patient Safety Agency (NPSA) identified EDS difficulties as a prominent safety issue in people with learning disability [143]. The LeDeR annual report highlighted EDS difficulties as one of the most common long-term conditions experienced by people with learning disability with 16% of deaths reported to LeDeR attributable to AP [6].

Early involvement of speech and language therapy services is paramount whenever there are concerns regarding swallowing difficulties in people with learning disability.

In these situations, it is important that there is a thorough structured review of medications associated with EDS difficulties e.g. antipsychotics [144,145]. The continued need for such medication

should be reviewed and reduction of dose or cessation considered. Notably, antipsychotics are prescribed more frequently for people with learning disability than the general population and often inappropriately [146].

The parallel BTS Aspiration Pneumonia statement covers the pathogenesis, investigation and management of EDS difficulties in detail [131].

Table 3 summarises the assessment, investigation and management of EDS difficulties.

Assessment and Investigation	Notes	Considerations
Clinical case history gathering	A careful history is important in the assessment of an individual's eating and drinking competence and in the identification of potential "risk factors" for EDS difficulties [147,148]. Typical features should be sought from the history including: • History of choking episodes • Coughing during and/or after meals • Increased shortness of breath when eating or drinking • Dysarthria • 'Bubbly' voice quality • Slow eating and/or refusing food • Regurgitation	Obtaining information from different sources, particularly family and carers, is vital to provide a more holistic picture of eating, drinking and swallowing abilities. Eating, drinking and swallowing abilities may change significantly over time.
Clinical swallow assessment (See pages 15-16 of parallel BTS Aspiration Pneumonia statement for further detail [131])	Clinical screening and swallow assessments are an essential starting point when evaluating a patient's eating and drinking safety and function [149]. Clinical assessments reliably identify EDS difficulties and overt signs of aspiration (e.g. choking, coughing and voice change). These assessments may inform decisions regarding further instrumental assessments (see below) and can be used to develop management plans and monitor progress [150].	The approach to swallow assessment varies widely [151–153]. There is a need for further evidence-based guidelines for the assessment of patients with oropharyngeal swallowing difficulties [149].
Instrumental assessment (See pages 15-16 of parallel BTS Aspiration Pneumonia statement for further detail [131]) These can include: • Videofluoroscopy of swallow (VFS) • Fibreoptic Endoscopic Evaluation of swallow (FEES)	A SLT is integral when considering the value and appropriateness of instrumental assessments. Decision making around the use of instrumental assessments should be undertaken by a multidisciplinary team (MDT), with the appropriate expertise.	If instrumental assessment is planned, careful consideration of the need for reasonable adjustments must be made. This assessment is a "snapshot" in time and does not account for variation of the persons swallow/ fatigue/ time of day/ positioning etc.

Management	Notes	Considerations
Compensatory strategies for managing risk in EDS difficulties. (See page 15 of parallel BTS Aspiration Pneumonia statement for further detail [131])	There are several compensatory strategies that may be utilised in the management of EDS difficulties (postural adjustments, diet modification, change of eating habits, feeding strategies). Limited evidence of impact on incidence of CAP.	Most appropriate strategy will depend on the stage of the swallow that has been affected (identified through careful history and examination). Use of compensatory strategies relies upon the individual or their family/carers being able to carry out the strategy consistently.
Enteral feeding (See pages 18-21 of parallel BTS Aspiration Pneumonia statement for further detail [131])	Should be considered if all forms of oral feeding are considered to result in a high risk of pneumonia. There is some weak evidence of reduced incidence of CAP following introduction of enteral feeding [145]. Important to consider other comorbidities (eg gastro-oesophageal reflux) when determining most appropriate enteral feeding method (gastrostomy, jejunostomy)	Careful risk/benefit discussions required with MDT, patient and family/carers. If not orally fed, patients may be able to take small tastes multiple times a day to encourage purposeful swallows and reduce posterior drooling.

Table 3 Assessment, investigation and management of eating, drinking and swallowing difficulties in people with learning disability

Clinical Practice Points

- Eating, drinking and swallowing (EDS) difficulties may be a contributing factor to CAP.
- Early involvement of speech and language specialists is essential for all those with a potential history of EDS difficulties.

Gastro-oesophageal reflux disease

Gastro-oesophageal reflux (GOR) is the reflux of gastric contents other than air into or through the oesophagus. Gastroesophageal reflux disease (GORD) is defined as a condition in which reflux leads to "troublesome symptoms and/or complications" [154]. GORD can result in several respiratory conditions, including recurrent AP [155]. Respiratory complications of GORD can result from acid, weakly acidic or non-acid reflux [156].

GORD has a higher incidence in people with learning disability than the general population. The prevalence of GORD has been reported between 31% to 75% in children with CP [157–159].

It can be difficult to recognise GORD in people with learning disability due to non-specific symptomatology and communication difficulties. Therefore, a high index of suspicion should be maintained when assessing for potential GORD. Typical symptoms such as heartburn, waterbrash and epigastric pain may not be determinable and, in those with severe learning disability, clinical features such as rumination, vomiting and haematemesis may predominate [160].

Objective measures, including 24-hour multichannel intraluminal impedance with single or dual channel pH measurement and endoscopy with oesophageal biopsy, are recommended for the diagnosis of GORD particularly in patients with neurodisability or where accurate evaluation of a significant response to proton pump inhibitor (PPI) therapy is not possible [161]. High resolution oesophageal manometry can be used for further elucidation of GORD pathophysiology particularly if there is clinical concern regarding possible oesophageal dysmotility[162]. Extraoesophageal biomarkers such as lipid laden macrophages and bronchoalveolar lavage pepsin are not diagnostically helpful [163]. In certain groups of patients with high prevalence of GORD (e.g. CP), a trial of PPIs with close monitoring of discernible clinical features may be the preferred diagnostic route.

Where GORD is deemed a potential cause of recurrent AP, referral to a specialist service should be considered for further investigation and management.

Table 4 outlines management options for GORD.

Treatment	Notes	Treatment Impact
Lifestyle modifications	Raising head of the bed, weight loss, avoiding recumbency following meals and having frequent small feeds are recommended for patients with GORD [163,164].	The impact of lifestyle modifications (either alone or in combination) on the incidence of pneumonia has not been determined.
PPIs	There is no evidence of superiority between PPIs so prescribing is solely based on availability, cost and patient age [163,164].	PPIs remain first line treatment for GORD with good evidence of symptom response [161,163,164]. There is no evidence for a reduced incidence of AP following PPI treatment. The possible link between PPI treatment and increased risk of pneumonia remains unproven due to the low quality of available evidence [165]. Regular review of the continued need for PPIs is essential.
Fundoplication	Laparoscopic Nissen fundoplication is considered the gold standard for surgical treatment of severe GORD [163]. Fundoplication is associated with postoperative complications in up to 45% of cases including bloating, retching and small bowel obstruction [163,166].	In both paediatric and adult studies, fundoplication shows a good symptomatic response rate [167,168]. Fundoplication can reduce the frequency of AP in a carefully selected patient group [163,164,169]. Other risk factors for pneumonia should be excluded/effectively treated before surgery is considered. Thorough preoperative evaluation (including oesophageal motility studies and MDT input) is essential.
Transpyloric / Jejunal Feeding	Jejunal tube feeding is a safe and effective means of enteral feeding when gastric feeding is insufficient to meet caloric needs [170]. Jejunal feeding must be continuous. Radiological intervention is required for reinsertion if dislodged.	Gastrojejunal feeding is associated with a similar reduction in the frequency of AP when compared to fundoplication in children with neurologic impairment [171,172]. A trial of nasojejunal feeds or, for children with an established gastrostomy, a trial of gastro-jejunal feeds via a gastrojejunostomy tube, may be useful to further establish the contribution of GORD to lung disease before definitive anti-reflux surgery is planned.

Table 4 Management options for GORD.

Clinical Practice Points

- GORD can be difficult to recognise in people with learning disability and may present with atypical symptoms such as rumination and haematemesis.
- Where GORD is deemed a potential cause of recurrent AP, referral to a specialist service should be considered for further investigation and management.
- Fundoplication or jejunal feeding may be considered in carefully selected patients with severe GORD and recurrent episodes of aspiration pneumonia where other risk factors have been excluded or optimally treated.

Excessive Oral Secretions

Sialorrhoea is an increase of saliva in the mouth. Sialorrhoea is common in individuals with neurodisability and most often occurs as the result of poor oral motor control and swallowing difficulties in individuals with neuromuscular conditions, rather than because of excess saliva production.

Sialorrhoea can be anterior or posterior. Drooling occurs when sialorrhoea is anterior and saliva spills out of the oral cavity. Posterior drooling is not visible and is more challenging to diagnose; it may accompany anterior drooling but can occur in isolation. In individuals with swallowing difficulties, posterior drooling causes saliva pooling in the oropharynx and hypopharynx. This creates a risk of aspiration and subsequent respiratory disease.

A history of frequent coughing, gagging, choking, chest congestion, or "wet" or gurgling sounds may indicate the presence of posterior drooling. Diagnosis of posterior drooling can be challenging; fibre-optic nasopharyngoscopy and laryngoscopy may allow direct visualisation of aspiration of secretions. In most cases, careful history and examination is adequate to guide the need for intervention.

Drooling can be assessed quantitatively with a variety of tools for severity and frequency (e.g. drooling quotient, number of bib changes) which can help evaluate response to interventions[173].

Management of sialorrhoea

Treatable causes of sialorrhoea include inadequately controlled GORD, nasal obstruction leading to open-mouthed posture, inadequate postural management, poor dental hygiene and medication side effects. If sialorrhoea continues despite optimisation of contributing factors, then specific sialorrhoea interventions should be considered. There is a paucity of evidence for management of sialorrhoea in adults with learning disability; most of the evidence relates to those with motor neuron disease and parkinson's disease. In CYP, sialorrhoea management has been studied most extensively in CP.

Table 5 outlines the most common interventions for sialorrhoea.

Intervention	Advantages	Disadvantages	Common side effects/cautions	Notes
Non-pharmacological interventions				
Behavioural interventions, oral motor therapy and oral appliances [174]	Non-invasive	Intensive Requires careful patient selection Weak evidence base for benefit[175]	Contra-indicated in posterior drooling[176].	
Medication				
Hyoscine transdermal patch	Patch change every 72 hrs Shown to be equally as efficacious as Glycopyrronium in treating sialorrhoea in CYP with CP [177].	Dose titration challenging Local skin irritation	Overly dry mouth, blurred vision, constipation, urinary retention, skin flushing or dryness.	Anticholinergic medications reduce saliva production either through direct or systemic route. Recommended as first line therapy for sialorrhoea in CYP and adults[176,178]. Can cause thick, difficult to clear secretions or mucous plugging resulting in CAP.
Glycopyrronium liquid	Dose titration Side effects less frequent than with Hyoscine hydrobromide[177].	Typical dosing schedule three times per day		
Inhaled Ipratropium bromide	Can be a useful adjunct to other medications listed. Less systemic absorption.	Facemask and spacer may be poorly tolerated Efficacy evidence base lacking.		
Atropine	Can be administered sublingually as drops No reliance on gut absorption [179].	Typical dosing 3-4 times per day Difficult to deliver accurate dosing.		
Salivary Gland Botulinum A Toxin injection	Often done under local anaesthetic with ultrasound guidance [180,181]. Can be repeated every 3-6 months	Invasive Repeated injections likely to be required	Trauma at injection site, EDS difficulties, dry mouth.	Cohort study evidence of reduced incidence of pneumonia following injection in CYP with neurologic impairment [182,183].
Surgical				
Salivary gland duct ligation	Results in the atrophy in salivary glands and subsequent reduction in saliva production [184].	Invasive Irreversible Requires general anaesthetic	Salivary gland stones	Some cohort study evidence of reduced incidence of CAP and hospitalisations in CYP with neurologic impairment [185,186].
Salivary gland excision	Highly effective [184].	Invasive Irreversible Requires general anaesthetic	External scarring Dry mouth Facial and hypoglossal nerve damage	

889 **Table 5** Sialorrhoea Interventions

Clinical Practice Points

- The identification of posterior drooling as a risk factor for CAP can be challenging, necessitating careful history and examination. Frequent coughing, gagging, choking, "wet"/gurgling sounds or daytime anterior drooling that ceases at night may be indicative.
- Optimisation of co-morbidities and other factors contributing to sialorrhoea may help improve saliva control. These include inadequately controlled GORD, nasal obstruction leading to open-mouthed posture, inadequate postural management, poor dental hygiene and medication side effects.
- First line pharmacological management comprises antimuscarinic drugs (hyoscine hydrobromide, glycopyrronium). Dose titration is necessary to avoid drying secretions excessively since lower airway secretions may become thicker and secretion clearance may become difficult.
- If first line pharmacological management is unsuccessful, salivary gland Botulinum A Toxin injection and surgical interventions should be considered in problematic sialorrhoea

Poorly controlled seizures

The prevalence of epilepsy in people with learning disability is significantly greater than in the general population (22.2% in a recent meta-analysis [187]). Seizures are associated with an increased risk of aspiration, including of vomit or saliva [38]. The risk of aspiration is further increased by the reduced muscle tone or drowsiness induced by medications used to control or terminate seizures [18]. National guidance should be employed for the optimal diagnosis, investigation and management of epilepsy [188].

Clinical Practice Points

- The diagnosis, investigation and management of epilepsy in people with learning disability should be optimised according to national guidance

Oral Health

Poor oral care and decaying teeth are risk factors for pneumonia in older people with some evidence in younger age groups [44,45]. People with learning disability experience more problems with their oral health than the general population [43].

The oral cavity is a complex microenvironment consisting of multiple bacterial and fungal species, their associated biofilms, and a cytokine milieu influenced by constant inflammatory stimulation [189]. Poor oral health promotes colonisation of the oral cavity with organisms such as *Haemophilus influenzae* and *Klebsiella pneumoniae*. It is postulated that micro aspiration of these organisms can initiate AP. The risk of AP appears to be greatest when these factors are compounded by chewing and swallowing difficulties [190].

In studies of older people and intensive care patients, oral health care consisting of tooth brushing at least twice daily and professional oral health care once a week reduces the incidence of AP [191,192]. Dental care was associated with decreased recurrence of severe pneumonia in a recent cohort study in children with neurologic impairment [193].

Without further research in this area, and given the relative simplicity and absence of identified risk, the best advice would appear to be to maintain good oral hygiene in people with learning disability to reduce the overall bacterial load. Routine oral care includes brushing the teeth, tongue, palate and gums with a soft brush at least twice a day with a fluoride toothpaste cleaning all tooth surfaces to remove plaque and food. At the end of two minutes brushing, toothpaste should be spat out without rinsing. In patients with eating, drinking and swallowing difficulties, non-foaming toothpaste should be used to reduce the risk of aspiration of the product.

People with learning disability often rely on others to perform their mouth care, so it is important that carers have the knowledge and skill to manage the person's oral care. An oral health assessment should be undertaken for each individual and an oral health care plan developed, ideally in consultation with the person's dentist. All children are now encouraged to have their first dental check by one year of age and being seen at this early stage helps parents to put preventative measures in place to maintain oral health. Many people with a learning disability can be seen in general dental practice but community dental services or special care dental services are available to help those who are unable to use general dental services. Accessible information may help the person with learning disability and their carers to maintain good oral health (ref [194] provides an excellent source of information with links to additional resources).

Clinical Practice Points

- Oral health should be regularly assessed in people with learning disability and an oral health care plan developed, ideally in consultation with the person's dentist.
- Good proactive oral care is essential and is most conveniently achieved by brushing teeth and gums with a soft toothbrush at least twice a day with a fluoride toothpaste using non-foaming toothpaste in those with swallowing difficulties to reduce aspiration risk.
- People with learning disability may be reliant on others to provide oral care. This must not be jeopardised if care settings change e.g. on admission to hospital.

Nutritional Considerations

Data from systematic review demonstrate that being underweight is associated with an increased risk of CAP [49]. Contributing factors include alteration to respiratory and diaphragmatic muscle structure and function leading to weakened cough and impaired airway clearance. Whilst systematic review data are inconsistent regarding the relative risk of pneumonia in overweight or obesity [49,195], comorbidities associated with obesity, e.g. reduced mobility, OSA, and GORD, may negatively impact chest health [50].

Underweight and obesity are more common in people with learning disability than in the general population [46–48]. Specific learning disability diagnoses are associated with an increased risk of obesity e.g. DS and PWS, whereas diagnoses such as autism can be associated with sensory feeding aspects leading to restricted diet and undernutrition.

Identification of underweight and overweight/obesity should be further assessed and managed by an appropriately skilled MDT, in accordance with current national guidance [196–199].

The appropriate frequency of weight and height monitoring should be determined according to individual circumstance and should be included in the AHC. Body Mass Index (BMI) should be calculated. BMI poorly reflects body composition in individuals with neurologic impairment [200,201]

due to alterations in body composition in some neurodisability conditions, and alternative measures (e.g. skinfold thickness) are recommended [161].

There is some evidence of efficacy for intensive individualised weight management interventions for overweight adults with learning disability, but these are costly and not widely available [202]. More readily accessible interventions (eg group weight loss programmes) may require the provision of social support, encouragement and role modelling by well-informed carers [202].

Management of underweight depends on aetiology and may include optimisation of the mealtime environment and routines, postural management and positioning, optimising parent/caregiver mealtime techniques, use of adaptive equipment, calorie supplementation and enteral feeding intervention.

Clinical Practice Points

- An appraisal of nutritional status, including measurement of height and weight, should be a fundamental part of the assessment of people with learning disability.
- The diagnosis, investigation and management of nutritional disorders in people with learning disability should be optimised according to national guidance.

Vaccination

Both influenza and pneumococcal vaccines have been demonstrated to significantly reduce hospitalisation rates for CAP across a wide range of populations [203–205]. Evidence suggests that vaccine coverage rates are lower [206] and potentially avoidable hospitalisations due to vaccine-preventable pneumonia increased in those with learning disability [207]. A patients' vaccine history should be reviewed at every opportunity.

In the UK, annual influenza vaccination has been available for all people with learning disability since 2014 but this has not led to an appreciable increase in uptake. Public Health England have provided guidance to support GP surgeries in making necessary arrangements to maximise influenza vaccination of people with learning disability. This guidance also contains a useful resource repository [208]. For those individuals with a learning disability with a needle phobia, influenza immunisation using the live-attenuated nasal spray preparation while less effective in adults, may be considered as a reasonable adjustment [209].

Learning disability is not currently a specific indication for pneumococcal vaccination although this is regularly reviewed by the Joint Committee on Vaccination and Immunisation (JCVI) [210]. Pneumococcal vaccination forms part of the routine childhood immunisation programme (Pneumococcal Conjugate Vaccine-13). Individuals with an incomplete or unreliable vaccine history should be considered unimmunised and specific guidance is available for appropriate immunisation in this situation [211]. The JCVI has defined clinical risk groups where further pneumococcal vaccination (Pneumococcal Polysaccharide Vaccine (PPV) -23) is recommended [212]. These include people with chronic respiratory disease including children at risk of aspiration due to neurological disease. PPV-23 is considered to provide long-term protection with a single dose unless the patient has asplenia, reduced splenic function or chronic renal disease when 5 yearly boosters are recommended.

Consent must be obtained before the administration of any vaccines [213] and for people with learning disability there must be an assessment of mental capacity and if necessary, a best interest decision made [214–216].

Clinical Practice Points

- Vaccine history should be reviewed at every opportunity.
- Annual influenza vaccination is recommended for all people with learning disability.
- Pneumococcal vaccination should be considered as specified by JCVI recommendations
- People with learning disability should be considered as high priority in all vaccination programmes for seasonal respiratory infections.

Smoking

In adults, tobacco smoking is a recognised risk factor for CAP [51] and Environmental Tobacco Smoke (ETS) exposure in childhood is associated with an increased risk of hospitalisation for CAP and increased severity of disease once hospitalised [52]. Smoking status should be included in an AHC [68] and a smoking/ETS exposure history taken when people with learning disability present with pneumonia [217]. Simple screening questions can be employed to identify those children at risk of ETS exposure [218].

Smoking cessation advice and referral onto smoking cessation services should be offered to all people with learning disability who are current smokers [217,219] and, where significant ETS exposure is identified, to their parents/carers.

Clinical Practice Points

- Every opportunity should be taken to review smoking and ETS exposure status.
- Smoking cessation advice and referral onto smoking cessation services should be offered to all people with learning disability who are current smokers and, where significant ETS exposure is identified, to their parents/carers.

7. Aetiology, Diagnosis, Investigation & Management of Community Acquired Pneumonia

Community acquired pneumonia is an extremely heterogenous disease with a wide range of presenting features particularly in people with learning disability. Detailed information gathering from all potential sources (patient, families, carers) is essential to enable accurate diagnosis and optimal management.

This management of CAP in people with learning disability is directed by national guidelines [217,220,221]. This section provides a precis of these guidelines highlighting specific considerations for people with learning disability.

Aetiology

In both adults and children with CAP, the commonest causative bacteria are *Streptococcus pneumoniae* and *Haemophilus influenzae* [217,222].

In people with learning disability presenting with CAP, further consideration should be made as to whether this pneumonia was triggered by an aspiration event and whether the oral cavity microbiome or colonisation may be contributing [223]. The emerging consensus is that community acquired AP is commonly polymicrobial and studies have identified a significant role of other causative bacteria such as gram negative bacteria (e.g. *Klebsiella spp., Enterobacter spp.* and *Escherichia Coli*) and *staphylococcus aureus* [a].

Viruses may also be the causative agent including influenza A, influenza B and, particularly in children, respiratory syncytial virus (RSV) [224–226]. Bacterial co-infection may occur in the context of a viral pneumonia [224,227] with commonest bacterial co-infections including S*taph. aureus* and *Strep. pneumoniae*.

Early Recognition

Early recognition of CAP can be challenging in people with learning disability. A high index of suspicion should be maintained to significant changes from normal that are recognised by parents/carers. Typical symptoms and signs, such as cough and fever, may be absent and more subtle features may predominate including reduction in oral fluid intake, reduced appetite, reduced mobility, altered behaviour or increased somnolence. Tools such as RESTORE2 may be utilised by carers to help identify soft signs of deterioration in health [228].

Clinical Practice Points

- Early recognition of CAP can be challenging in people with learning disability. Soft signs, such as changes to baseline alertness and mobility, should be viewed with a high index of suspicion.

[a] Further details regarding the microbiology of Aspiration Pneumonia can be found on page 12 of the Aspiration Pneumonia Statement

Diagnosis

Symptoms and Signs

Symptoms that are typically associated with CAP include cough, history of fever, rapid breathing and breathlessness [217,220]. Symptoms vary considerably with age - young children may present with intermittent apnoea and grunting respiration whilst chest pain may be the predominant symptom in teenagers and adults [217,220,229]. A detailed clinical history should include enquiry about swallowing efficiency, recent choking on medication/food/liquids, neurological and gastrointestinal symptoms, the frequency of previous pneumonia and risk factors for CAP.

Typical signs of CAP include increased work of breathing, tachypnoea, fever, dullness to percussion and crackles or bronchial breathing on auscultation [217,220]. The incidence and nature of individual signs are age dependent. For example, head bobbing is a sign of increased work of breathing observed specifically in infancy [220].

Clinical features of CAP in individuals with learning disability should be appraised with appropriate consideration of individual circumstances and any associated comorbidities. A clinical history should be sought from the patient using communication aids as necessary (see Section 2: Engagement and Assessment). Parents and carers should be consulted for further detail and may serve as the primary source of information where direct communication with the patient is not possible.

Clinical examination should be adapted to account for associated comorbidities which may impact potential signs of CAP and, if available, details of the patient's baseline clinical status for comparison can be helpful. For example, scoliosis *per se* may result in features of CAP including tachypnoea, increased work of breathing and asymmetrical chest findings such as reduced breath sounds on the concave side [230].

Assessment should include consideration of the clinical features of risk factors for CAP in people with learning disability (see Section 2, Table 1).

Assessing severity

CAP severity in CYP with learning disability is determined by clinical assessment of symptoms and signs [220]. There are currently no validated severity assessment tools for CAP in children. Table 6 details clinical features that help define the severity of CAP. In addition to assessing severity, decisions regarding admission to hospital and management should take account of any underlying risk factors together with the ability of the parents/carers to manage the illness in the community.

	Mild to Moderate	Severe
Children Under 5 years age	Respiratory rate < 70 bpm under 12 months age, <50 bpm 12 months to 5 years Mild recession Taking full feeds	Tachypnoea (>70 bpm under 12 months age, >50 bpm 12 months to 5 years) Moderate/severe recession (<12 months) Severe difficulty breathing (>12 months) Grunting Nasal Flaring Apnoea (<12 months) Cyanosis Tachycardia Capillary Refill Time ≥ 2 secs Hypoxaemia (sustained oxygen saturation <92% in room air) Not feeding (< 12 months) Signs of dehydration (>12 months)
Older children (>5 years) and young people	Respiratory rate <50 breaths/min Mild breathlessness No vomiting	Tachypnoea (RR >50 bpm) Severe difficulty breathing Grunting Nasal Flaring Cyanosis Tachycardia Capillary Refill Time ≥ 2 secs Hypoxaemia (sustained oxygen saturation <92% in room air) Signs of dehydration

Table 6 Severity assessment in CYP (adapted with permission from BTS Guidelines for the Management of Community Acquired Pneumonia in Children[220])

In adults with learning disability and CAP, the CURB-65 score should be used to assess severity in conjunction with clinical judgement [217]. In patients being assessed in the community setting, the CRB-65 score should be used. Although these scoring systems have not been validated specifically for adults with learning disability or neurodisability, they have been validated within the general adult population and demonstrated to be good predictors of mortality in CAP [231]. Patients with a low risk score (0-1) have a low risk of mortality and can be managed safely in the community, unless there are alternative reasons for admission to hospital, such as social concerns. Patients with a moderate (2) or high risk (3-5) score should be admitted to hospital for management of CAP. Further investigation and treatment in adults with CAP will also be guided by their CURB-65/ CRB-65 score (Figure 4).

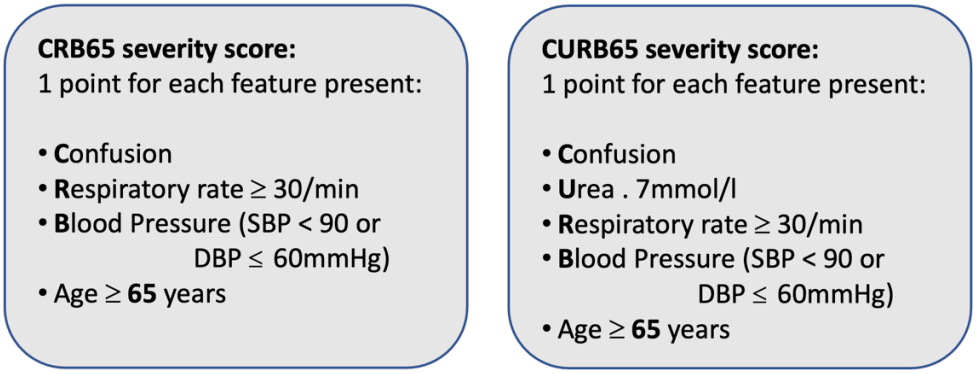

Figure 4 CRB-65 Score and CURB-65 Score

If the patient is identified at high risk of mortality, or fails to respond to initial treatment, the clinical team must initiate honest conversations with the patient and their family carers, explaining the possibility of death and exploring their wishes and priorities. Even if full active intervention is being pursued it is still important to warn loved ones of a possible poor outcome.

Clinical Practice Points

- A comprehensive history should be sought from the patient, using communication aids when required, with additional information gathering from parents and carers.
- The severity of CAP should be assessed to guide treatment and decision making around requirement for hospital assessment and admission. In adults with learning disability, the CRB-65 or CURB-65 should be used in conjunction with clinical judgement. In CYP, clinical features such as respiratory rate, difficulty in breathing and hydration status can be used to assess severity.

Investigation

Radiological Investigations

A chest radiograph can potentially provide valuable information in individuals with suspected CAP including the detection of consolidation and effusions. Interpretation may be difficult due to comorbidities such as reduced diaphragm excursion in neuromuscular disorders, under-expansion of the lungs in thoracic deformity and distortion of anatomical relationships in scoliosis.

In the community setting, it is not necessary to perform a chest radiograph unless the diagnosis is in doubt, there is a lack of clinical progress on treatment for suspected CAP, or if the patient is likely to have an underlying pathology [217,220].

In adult patients with learning disability, a chest radiograph should be obtained in all patients admitted to hospital [217]. A chest radiograph is not considered routine in CYP with learning disability admitted to hospital with CAP [220] but should be considered in those with features of severe pneumonia (particularly if hypoxic or with evidence of significant respiratory distress) or where there is clinical suspicion of a complicated pneumonia (absent breath sounds, dull to percussion, decreased chest expansion).

Chest radiography is part of the diagnostic workup in cases of suspected AP [b].

Microbiological Investigations

The routine collection of respiratory specimens or blood for microbiological analysis is not indicated in the majority of individuals with CAP [217,220].

In children, the collection of specimens for analysis should be considered in those with severe CAP and in those patients with a history of recurrent/frequent CAP or previous infection with MRSA or PA. Specimens should include nasopharyngeal and/or nasal swabs for viral PCR, sputum for culture and blood for culture and serology for respiratory viruses, Mycoplasma and Chlamydia [220]. Tracheal suction aspirates should be sent for culture in patients with a tracheostomy. Urinary antigen detection may help as a negative predictor of pneumococcal infection in older children but positive tests are non-specific and may represent carriage [220].

In adults with learning disability, investigation should be guided by their CURB-65 score. Further microbiological investigation is indicated in all patients presenting with CAP and a moderate or high risk CURB score (score of 2 or greater) [217]. Investigation should include collection of blood and sputum specimens for culture, prior to initiation of antibiotic therapy where possible, and pneumococcal urine antigen testing. *Legionella* pneumonia should be looked for in the context of high severity pneumonia, during an outbreak or if the patient has other specific risk factors. In patients with high severity CAP who are not responsive to ß-lactam antibiotics or with a strong clinical, epidemiological or radiological suspicion of 'atypical' pneumonia, further investigation should be considered for *Mycoplasma pneumoniae*, Chlamydia spp. and *Pneumocystis jirovecii* (if risk factors are present). Molecular testing of upper respiratory tract samples should also be considered as part of microbiological investigation [232].

In adults presenting with CAP, HIV testing should be considered [233], with CAP a recognised 'indicator illness'.

General Investigations

For patients who are managed in the community, the assessment of oxygenation by pulse oximetry can be a useful adjunct to clinical decisions regarding site of care and need for further investigation [217,220]. It is important to appreciate the potential limitations and technical issues associated with oximetry in certain groups of patients (e.g. those with movement disorders) [234]. Blood investigations are not generally indicated in the community setting [217,220].

In hospitalised children with learning disability, white blood cell (WBC) count, C-reactive protein (CRP), and procalcitonin are not routinely indicated as they are not clinically useful in predicting disease severity or outcome and cannot reliably differentiate viral from bacterial infection [235]. Pulse oximetry is an essential component of the diagnostic workup and subsequent management [220].

[b] The diagnosis of Aspiration Pneumonia is covered in detail on pages 23-24 of the Aspiration Pneumonia Statement

Adults with learning disability being assessed in hospital for CAP should have pulse, blood pressure, temperature, respiratory rate, pulse oximetry and level of consciousness measured and an appropriate early warning score calculated (e.g. NEWS2 or an equivalent scoring system) as part of the assessment [236]. This is particularly relevant in adult patients with learning disability, where a delay in recognition of clinical deterioration has been identified as a potentially avoidable contributory factor to deaths [6]. Full blood count, CRP, urea and electrolytes and liver function tests should also be performed [217]. An arterial blood gas or ear lobe blood gas should be performed if the patients is at risk of hypercapnic respiratory failure, including from obesity, kyphoscoliosis, neuromuscular weakness, or underlying COPD, and requires supplementary oxygen, in keeping with current national oxygen guidelines [237]. If an arterial blood gas is required then local anaesthetic should be used [237]. Patients being admitted should also undergo a venothromboembolism risk assessment, using a validated tool and low molecular weight heparin should be prescribed in patients unless the risk is felt to outweigh the benefit [217,238].

Clinical Practice Points

- A chest radiograph should be performed in all adults requiring hospital assessment with suspected CAP and should be considered in CYP with features of severe or complicated pneumonia.
- In CYP with severe CAP and adults with moderate to severe CAP (for example CURB 65>2), blood testing and microbiological investigations including sputum culture and sensitivity, blood cultures and pneumococcal urine antigen should be undertaken to help guide treatment.
- In all CYP and adults being assessed for suspected CAP, pulse oximetry should be undertaken as it can be a useful adjunct to clinical decisions regarding site of care and need for further investigation.
- For hospitalised adults with CAP, NEWS2 (or an equivalent early warning score) should be calculated and tracked throughout hospital assessment and admission to monitor for signs of deterioration and to facilitate timely clinical response.

Management

Antibiotic management

All CYP with learning disability who have a clear clinical diagnosis of pneumonia should receive antibiotics as bacterial and viral pneumonia cannot be reliably distinguished from each other [220]. Antibiotics administered orally/enterally are a safe and effective treatment for CYP presenting with even severe CAP [239]. Intravenous antibiotics should be used when the patient is unable to tolerate oral/enteral medicines (e.g. due to vomiting) or presents with features of septicaemia or complicated pneumonia. Antibiotics should be prescribed according to local and national guidelines [221]. Microbiological results may guide alternative antibiotic choice after consultation with local microbiology services.

In adults with learning disability, presenting with CAP to hospital, risk stratification using the CURB-65 in hospital settings or the CRB-65 in community settings should be undertaken [240,241] in conjunction with clinical judgement (Table 6):

Low risk CURB-65 0-1, CRB-65 0	Patients with a low-risk score have a low mortality and should be considered for home management, in conjunction with clinical judgement and review of social circumstances and comorbidities. Oral / enteral antibiotics should be prescribed according to local and national guidelines [221].
Moderate risk CURB-65 2, CRB-65 1-2	Patients with a moderate-risk score should be admitted to hospital. Treatment with oral / enteral antibiotics is the preferred option, according to local and national guidelines[221].
High risk CURB-65 3-5, CRB-65 3-4	Patients with a high-risk score should be admitted to hospital. Treatment with intravenous antibiotics is recommended according to local and national guidelines[221]

Table 6 Risk stratification using CURB-65 in hospital settings or CRB-65 in community settings. Should be employed in conjunction with clinical judgement.

Antibiotic choice should be guided by clinical severity, known pathogens and allergies. Antibiotics should be initiated as soon as possible, within 4 hours [221] of confirming the diagnosis of CAP, or within 1 hour if clinical signs of sepsis . If treated with intravenous antibiotics, this should be reviewed within 48 hours and consideration made to switch to oral antibiotics[221]. Antibiotics should be stopped after 5 days, unless the patient is clinically unstable or microbiological investigations suggest that a longer course is required[221].

The recommendations for treatment of aspiration pneumonia in patients with learning disability is that described in the parallel clinical statement “Aspiration Pneumonia”.

Oxygen

All patients admitted to hospital with CAP should have their oxygenation checked.

CYP with learning disability who have been admitted to hospital should be treated with oxygen if their oxygen saturation is <92% in air[220]. Oxygen should be administered via nasal cannulae or face mask to maintain oxygen saturation 93-98%. An increasing body of evidence supports the use of high flow nasal cannula (HFNC) oxygen in cases with significant respiratory distress [242].

In adults with learning disability presenting with CAP, oxygen should be prescribed to a target oxygen saturation of 94-98% unless at risk of hypercapnic respiratory failure [237]. Oxygen should be administered in keeping with BTS Emergency Oxygen guidelines [237]. More conservative oxygen therapy with lower target saturations is increasingly employed and an upper target of 94-96% may be optimal [243]. Patients with learning disability may be at high risk of hypercapnic respiratory failure due to underlying COPD, obesity, scoliosis or neuromuscular weakness. In this group of patients, target saturations should be 88-92% [237].

There is a role for HFNC oxygen in adults with learning disability presenting with CAP who have acute hypoxic respiratory failure. HFNC therapy is better tolerated than conventional face mask oxygen [244]. The heated humidification is beneficial for mucociliary clearance [245] as compared to conventional oxygen therapy which can be drying to the mouth and to respiratory secretions. In acute hypoxic respiratory failure, it has been demonstrated to be both safe and effective [246].

Airway clearance

In most cases of pneumonia, lung tissue consolidation exists without excess secretions. In this situation there is no evidence that respiratory physiotherapy is of benefit [247]. The BTS guidelines state patients with pneumonia should not be routinely treated with ACTs [220,240,248]. However, where retention of secretions is present or a patient has recurrent excess secretions ACTs are indicated [240,248] and early referral for chest physiotherapy should be considered.

Clinical Practice Points

- Antibiotic therapy should be guided by clinical severity, known pathogens and allergies.
- In CYP with CAP, supplementary oxygen should be administered to maintain oxygen saturations 93-98%.
- In adults, target oxygen saturations between 94-98% should be used in people where there is no risk of hypercapnic respiratory failure. For those with risk factors for hypercapnic respiratory failure (such as scoliosis, neuromuscular disease, COPD or obesity), oxygen saturations should be maintained between 88-92%.
- In patients with retention of secretions, early involvement of chest physiotherapy should be considered.

Escalation of care

Patients with learning disability admitted to hospital with CAP who have signs of sepsis or acute hypoxic or hypercapnic respiratory failure should have early involvement of critical care services. The LeDeR report [6] has highlighted the need for early involvement and a lower threshold for admission to critical care for CYP with learning disability. Although there is little evidence to help guide decision making for adults with learning disability requiring critical care, patients with learning disability should have the same access to critical care services as other service users [249].

Care should be taken to avoid diagnostic overshadowing, defined as "symptoms of physical ill health mistakenly attributed to either a mental health/behavioural problem or as being inherent in the person's learning disabilities" [250]. Such diagnostic over-shadowing may prevent a clinician recognising a patient's deterioration or delay initiation of key treatments or discussions with critical care services regarding escalation to HDU or ICU.

It is important to remember that patients with profound or multiple learning disability, at presentation with a severe intercurrent illness, can neurologically deteriorate which can result in clinicians making inappropriate assumptions about quality of life at baseline. In addition, making a judgement on a patient's quality of life without full assessment risks providing inadequate, impersonal care. Where a patient is unable to communicate their quality of life, proxies are almost as good at detecting changes in quality of life as the individual [251]. Any advance directive (or advance care plan) by the patient should also be considered along with previous discussions around appropriateness of escalation between clinicians, the patient and their family. The clinical frailty score should not be used to make escalation decisions for people with learning disability or long term stable disability as it has not been validated in these groups [13].

There are no studies to suggest that patients with learning disability requiring acute ventilatory support for pneumonia have adverse outcomes on critical care compared to other patient groups. Importantly, studies in children with neurodisability requiring paediatric critical care did not show any association between baseline functional status and outcome from critical care admission [252]. Decisions should be made on whether intubation and ventilation or other forms of ventilatory

support are appropriate according to the trajectory of the patient in conjunction with relevant comorbidities. If a patient, including those with profound or multiple disabilities, has a stable trajectory then they should not be excluded from access to critical care or from invasive ventilation [253]. Discussions around ventilation should involve the patient, if they have capacity to be involved in decision making, or proxy where possible.

It is essential that adequate time is given to discuss treatment expectations around escalation. The views of the patient and their advocates (parents/carers) should be fully explored, often as part of advance care planning. This can reduce the risk of significant disagreement which may negatively impact trust between staff and patients and their advocates. Strategies to promote an effective relationship between healthcare professionals and people with learning disability and their advocates include:

- learning disability training for all healthcare staff [254] to promote reasonable adjustments and avoid potential discrimination or assumptions about quality of life
- early identification of patients with a learning disability (e.g. automatic alerts on electronic patient records) to enable early referral to learning disability liaison teams and proactive care
- recognition of the expertise held by parents and carers, and valuing their role as a vital part of the MDT
- regular transparent MDT discussion to review progress and goals and for care responsibilities to be negotiated and agreed

In the rare cases where conflict does occur, steps should be taken to maintain effective dialogue between parties. Failure to recognise conflict and resolve it an early opportunity may lead to court action or public confrontation [255]. Offering a second opinion and/or requesting the opinion of a local clinical ethics committee can support the patient, their advocates and health professionals to come to the best decision for the patient. Formal medical mediation may also be valuable [256].

Previous LeDeR reports have highlighted the inappropriate use of Do Not Attempt Cardiopulmonary Resuscitation (DNACPR) documents in patients with learning disability, with 'DNACPR' forms completed incorrectly with learning disability as the sole reason to not attempt resuscitation [6]. Discussions around decisions about cardiopulmonary resuscitation must be clearly discussed with the patient or family/carer [257].

Clinical Practice Points

- In patients with signs of sepsis, acute hypoxic or hypercapnic respiratory failure, early involvement of critical care services should be undertaken.
- Decisions around escalation to critical care should involve patients, families and carers along with clinical teams who are involved in the delivery of routine clinical care to the patient.
- Decisions around escalation of care should take into consideration the trajectory of the patient prior to admission.
- The clinical frailty score should not be used as part of decision making in patients with learning disability presenting with CAP.

Consideration of CAP risk factors and follow up

An episode of CAP indicates an increased risk of subsequent CAP [26] particularly in patients with CP [27,28]. It is paramount that the opportunity is taken to make a full respiratory assessment, review the possible contribution of risk factors and arrange follow up, further investigation and management as necessary. Where a deterioration in an underlying degenerative condition has been identified, the ongoing risk of further episodes should be acknowledged. This is an opportunity for advance care planning to take place to ensure the patient's wishes for future treatment can be clearly communicated to clinicians responding to a future event.

Follow up should be organised according to radiological findings, the severity of CAP and need for further evaluation of risk factors.

CYP hospitalised with severe pneumonia, empyema or lung abscess should be followed up after discharge until they have recovered completely and their chest x-ray has returned to near normal [220].

For adult patients presenting with CAP, follow up in a respiratory clinic or primary care is recommended at 6 weeks [217]. A follow up chest radiograph at 6 weeks should be undertaken for those at risk of underlying malignancy such as those with a significant smoking history or those over the age of 50 years [240].

All patients who present with CAP and have been identified as having risk factors should have a follow up arranged so that a holistic assessment of the patient's needs can be made, and risk factors addressed. (See section 1).

Clinical Practice Points

- An episode of CAP should trigger a full respiratory assessment including detailed clinical history and examination, review of risk factors and consideration for pertinent investigations.
- Follow up should be arranged for all patients who present with CAP and have been identified as having risk factors so that a holistic assessment of the patient's needs can be made, and risk factors addressed.

8. Palliative Care Considerations

This section may be relevant for patients with learning disability who fit into any of the following categories:

- Patients already known to be reaching the end of life who develop pneumonia either as a direct consequence of their underlying condition, or due to their increasing frailty.
- Patients who may have had multiple episodes of pneumonia, where there is a deteriorating trajectory and there is concern that their quality of life is getting worse.
- Patients who have a significant symptom burden or who appear to be sick enough to die because of their pneumonia, even if there is uncertainty and active treatment is still being pursued.

Many of the patients who fit into these groups may subsequently recover partially or completely. In these patients taking time to consider a palliative approach should not be considered to conflict with active treatment.

Definitions

End of life: Patients are 'approaching the end of life' when they are likely to die within the next 12 months. This includes patients whose death is imminent (expected within a few hours or days). In practice, the term 'end of life' is interpreted in different ways but usually refers to the final phase of illness.

Palliative care: Palliative care is an approach that improves the quality of life of patients (adults and children) and their families/carers who are facing problems associated with life-threatening illness. It prevents and relieves suffering through the early identification, correct assessment and treatment of pain and other problems, whether physical, psychosocial or spiritual (World Health Organization definition).

Palliative care (encompassing end-of-life care and a period of possibly weeks/months/years earlier) may be useful for patients with advanced progressive illness, even if there is still hope of cure [258]. There has been very little research looking at the palliative care needs of those with learning disability [259,260]. Early referral to palliative care teams is recommended specifically for those with learning disability [10,259,260].

A specialist palliative care team will be able to assess and manage physical, psychological and spiritual symptoms to reduce suffering and distress. They can also offer valuable input into complex clinical decision-making challenges and support the people around the patient. There is a potential role for specialist palliative care teams with expertise in learning disability, or additional training in end-of-life care for learning disability nurses [261].

The 'surprise' question is a simple test when considering whether a patient is likely to benefit from palliative care. Health professionals can ask themselves "Would I be surprised if this patient were to die within the next 12 months?". If the answer is no, this provides recognition that a patient may have transitioned into a new phase of illness [262]. Palliative care can be delivered alongside active treatment when appropriate (parallel planning).

Appendix 2 provides further detail regarding relevant palliative care considerations including guidance on breaking bad news, legal aspects in decision making, advance care plans, treatment of infections in end of life (EOL), clinically assisted nutrition and hydration in EOL, symptom management, preferred place of care, and bereavement support for people with learning disability.

Clinical Practice Points

- All health professionals should be able to provide quality palliative care with support from specialist palliative care teams where necessary.
- Palliative care can be delivered alongside active treatment in a parallel planning approach. The two are not mutually exclusive.
- Palliative care focusses on quality of life. Developing a personalised plan in advance can help to record a patient's usual quality of life, priorities and goals. The patient should be supported to contribute to this as much as they are able along with family or friends who can advocate for them.

9. References

(1) Department of Health and Social Care. Valuing People: A New Strategy for Learning Disability for the 21st Century. London: The Stationary Office 2001.

(2) National Institute for Health and Care Excellence. Learning Disabilities: Background Information: Definition. London 2020.

(3) Public Health England. People with Learning Disabilities in England 2015: Main Report. London, UK 2016.

(4) NHS Digital. Quality and Outcomes Framework, 2020-21 https://digital.nhs.uk/data-and-information/publications/statistical/quality-and-outcomes-framework-achievement-prevalence-and-exceptions-data/2020-21 (accessed 2022 -02 -16).

(5) NHSE. NHSEI 2019. Improving Identification of People with a Learning Disability: Guidance for General Practice. **2019**.

(6) Learning Disabilities Mortality Review (LeDeR) Programme (2019) Annual Report. Bristol: Norah Fry Centre for Disability Studies, University of Bristol. December 2017.

(7) Glover, G.; Williams, R.; Oyinlola, J. An Observational Cohort Study of Numbers and Causes of Preventable General Hospital Admissions in People with and without Intellectual Disabilities in England. *J. Intellect. Disabil. Res.* **2020**, *64* (5), 331–344. https://doi.org/10.1111/jir.12722.

(8) Chang, C.-K.; Chen, C.-Y.; Broadbent, M.; Stewart, R.; O'Hara, J. Hospital Admissions for Respiratory System Diseases in Adults with Intellectual Disabilities in Southeast London: A Register-Based Cohort Study. *BMJ Open* **2017**, *7* (3), e014846. https://doi.org/10.1136/bmjopen-2016-014846.

(9) LeDeR team. LeDeR Action from Learning Report 2020/21. University of Bristol: Norah Fry Centre for Disability Studies. 2021.

(10) Heslop, P.; Blair, P. S.; Fleming, P.; Hoghton, M.; Marriott, A.; Russ, L. The Confidential Inquiry into Premature Deaths of People with Intellectual Disabilities in the UK. 2013.

(11) Tuffrey-Wijne, I.; Giatras, N.; Goulding, L.; Abraham, E.; Fenwick, L.; Edwards, C.; Hollins, S. *Identifying the Factors Affecting the Implementation of Strategies to Promote a Safer Environment for Patients with Learning Disabilities in NHS Hospitals: A Mixed-Methods Study*; Health Services and Delivery Research; NIHR Journals Library: Southampton (UK), 2013.

(12) Cluley, V. From "Learning Disability to Intellectual Disability"—Perceptions of the Increasing Use of the Term "Intellectual Disability" in Learning Disability Policy, Research and Practice. *Br. J. Learn. Disabil.* **2018**, *46* (1), 24–32. https://doi.org/10.1111/bld.12209.

(13) National Institute for Health and Care Excellence. COVID-19 rapid guideline: managing COVID-19 https://www.nice.org.uk/guidance/ng191 (accessed 2021 -08 -20).

(14) Kinnear, D.; Morrison, J.; Allan, L.; Henderson, A.; Smiley, E.; Cooper, S.-A. Prevalence of Physical Conditions and Multimorbidity in a Cohort of Adults with Intellectual Disabilities with and without Down Syndrome: Cross-Sectional Study. *BMJ Open* **2018**, *8* (2), e018292. https://doi.org/10.1136/bmjopen-2017-018292.

(15) Peristeri, E.; Aloizou, A.-M.; Keramida, P.; Tsouris, Z.; Siokas, V.; Mentis, A.-F. A.; Dardiotis, E. Cognitive Deficits in Myopathies. *Int. J. Mol. Sci.* **2020**, *21* (11), 3795. https://doi.org/10.3390/ijms21113795.

(16) Towards a definition of neurodisability: a Delphi survey - Morris - 2013 - Developmental Medicine & Child Neurology - Wiley Online Library https://onlinelibrary.wiley.com/doi/full/10.1111/dmcn.12218 (accessed 2021 -08 -14).

(17) Marpole, R.; Blackmore, A. M.; Gibson, N.; Cooper, M. S.; Langdon, K.; Wilson, A. C. Evaluation and Management of Respiratory Illness in Children With Cerebral Palsy. *Front. Pediatr.* **2020**, *8*, 333. https://doi.org/10.3389/fped.2020.00333.

(18) Gibson, N.; Blackmore, A. M.; Chang, A. B.; Cooper, M. S.; Jaffe, A.; Kong, W.; Langdon, K.; Moshovis, L.; Pavleski, K.; Wilson, A. C. Prevention and Management of Respiratory Disease in Young People with Cerebral Palsy: Consensus Statement. *Dev. Med. Child Neurol.* **2021**, *63* (2), 172–182. https://doi.org/10.1111/dmcn.14640.

(19) Hull, J.; Aniapravan, R.; Chan, E.; Chatwin, M.; Forton, J.; Gallagher, J.; Gibson, N.; Gordon, J.; Hughes, I.; McCulloch, R.; Russell, R. R.; Simonds, A. British Thoracic Society Guideline for Respiratory Management of Children with Neuromuscular Weakness. *Thorax* **2012**, *67* (Suppl 1), i1–i40. https://doi.org/10.1136/thoraxjnl-2012-201964.

(20) Birnkrant, D. J.; Bushby, K.; Bann, C. M.; Alman, B. A.; Apkon, S. D.; Blackwell, A.; Case, L. E.; Cripe, L.; Hadjiyannakis, S.; Olson, A. K.; Sheehan, D. W.; Bolen, J.; Weber, D. R.; Ward, L. M.; DMD Care Considerations Working Group. Diagnosis and Management of Duchenne Muscular Dystrophy, Part 2: Respiratory, Cardiac, Bone Health, and Orthopaedic Management. *Lancet Neurol.* **2018**, *17* (4), 347–361. https://doi.org/10.1016/S1474-4422(18)30025-5.

(21) Byrne, J. H.; Lennox, N. G.; Ware, R. S. Systematic Review and Meta-Analysis of Primary Healthcare Interventions on Health Actions in People with Intellectual Disability. *J. Intellect. Dev. Disabil.* **2016**, *41* (1), 66–74. https://doi.org/10.3109/13668250.2015.1105939.

(22) Goldbart, A. D.; Tal, A.; Givon-Lavi, N.; Bar-Ziv, J.; Dagan, R.; Greenberg, D. Sleep-Disordered Breathing Is a Risk Factor for Community-Acquired Alveolar Pneumonia in Early Childhood. *Chest* **2012**, *141* (5), 1210–1215. https://doi.org/10.1378/chest.11-1998.

(23) Chiner, E.; Llombart, M.; Valls, J.; Pastor, E.; Sancho-Chust, J. N.; Andreu, A. L.; Sánchez-de-la-Torre, M.; Barbé, F. Association between Obstructive Sleep Apnea and Community-Acquired Pneumonia. *PLOS ONE* **2016**, *11* (4), e0152749. https://doi.org/10.1371/journal.pone.0152749.

(24) Bach, J. R.; Rajaraman, R.; Ballanger, F.; Tzeng, A. C.; Ishikawa, Y.; Kulessa, R.; Bansal, T. Neuromuscular Ventilatory Insufficiency: Effect of Home Mechanical Ventilator Use v Oxygen Therapy on Pneumonia and Hospitalization Rates. *Am. J. Phys. Med. Rehabil.* **1998**, *77* (1), 8–19. https://doi.org/10.1097/00002060-199801000-00003.

(25) Tzeng, A. C.; Bach, J. R. Prevention of Pulmonary Morbidity for Patients with Neuromuscular Disease. *Chest* **2000**, *118* (5), 1390–1396. https://doi.org/10.1378/chest.118.5.1390.

(26) Almirall, J.; Serra-Prat, M.; Bolíbar, I.; Balasso, V. Risk Factors for Community-Acquired Pneumonia in Adults: A Systematic Review of Observational Studies. *Respiration* **2017**, *94* (3), 299–311. https://doi.org/10.1159/000479089.

(27) Blackmore, A. M.; Bear, N.; Blair, E.; Langdon, K.; Moshovis, L.; Steer, K.; Wilson, A. C. Predicting Respiratory Hospital Admissions in Young People with Cerebral Palsy. *Arch. Dis. Child.* **2018**, *103* (12), 1119–1124. https://doi.org/10.1136/archdischild-2017-314346.

(28) Meehan, E.; Freed, G. L.; Reid, S. M.; Williams, K.; Sewell, J. R.; Rawicki, B.; Reddihough, D. S. Tertiary Paediatric Hospital Admissions in Children and Young People with Cerebral Palsy. *Child Care Health Dev.* **2015**, *41* (6), 928–937. https://doi.org/10.1111/cch.12263.

(29) Blair, E.; Watson, L.; Badawi, N.; Stanley, F. J. Life Expectancy among People with Cerebral Palsy in Western Australia. *Dev. Med. Child Neurol.* **2001**, *43* (8), 508–515. https://doi.org/10.1017/s0012162201000949.

(30) Hilgenkamp, T. I. M.; Reis, D.; van Wijck, R.; Evenhuis, H. M. Physical Activity Levels in Older Adults with Intellectual Disabilities Are Extremely Low. *Res. Dev. Disabil.* **2012**, *33* (2), 477–483. https://doi.org/10.1016/j.ridd.2011.10.011.

(31) Carmeli, E.; Merrick, J.; Kessel, S.; Masharawi, Y.; Carmeli, V. Elderly Persons with Intellectual Disability: A Study of Clinical Characteristics, Functional Status, and Sensory Capacity. *ScientificWorldJournal* **2003**, *3*, 298–307. https://doi.org/10.1100/tsw.2003.24.

(32) Ball, S. L.; Panter, S. G.; Redley, M.; Proctor, C.-A.; Byrne, K.; Clare, I. C. H.; Holland, A. J. The Extent and Nature of Need for Mealtime Support among Adults with Intellectual Disabilities. *J. Intellect. Disabil. Res. JIDR* **2012**, *56* (4), 382–401. https://doi.org/10.1111/j.1365-2788.2011.01488.x.

(33) Chadwick, D. D.; Jolliffe, J. A Descriptive Investigation of Dysphagia in Adults with Intellectual Disabilities. *J. Intellect. Disabil. Res.* **2009**, *53* (1), 29–43. https://doi.org/10.1111/j.1365-2788.2008.01115.x.

(34) Robertson, J.; Chadwick, D.; Baines, S.; Emerson, E.; Hatton, C. Prevalence of Dysphagia in People With Intellectual Disability: A Systematic Review. *Intellect. Dev. Disabil.* **2017**, *55* (6), 377–391. https://doi.org/10.1352/1934-9556-55.6.377.

(35) Robertson, J.; Chadwick, D.; Baines, S.; Emerson, E.; Hatton, C. People with Intellectual Disabilities and Dysphagia. *Disabil. Rehabil.* **2018**, *40* (11), 1345–1360. https://doi.org/10.1080/09638288.2017.1297497.

(36) Hsu, W.-T.; Lai, C.-C.; Wang, Y.-H.; Tseng, P.-H.; Wang, K.; Wang, C.-Y.; Chen, L. Risk of Pneumonia in Patients with Gastroesophageal Reflux Disease: A Population-Based Cohort Study. *PLoS ONE* **2017**, *12* (8), e0183808. https://doi.org/10.1371/journal.pone.0183808.

(37) Erasmus, C. E.; van Hulst, K.; Rotteveel, J. J.; Willemsen, M. A. A. P.; Jongerius, P. H. Clinical Practice: Swallowing Problems in Cerebral Palsy. *Eur. J. Pediatr.* **2012**, *171* (3), 409–414. https://doi.org/10.1007/s00431-011-1570-y.

(38) DeToledo, J. C.; Lowe, M. R.; Gonzalez, J.; Haddad, H. Risk of Aspiration Pneumonia after an Epileptic Seizure: A Retrospective Analysis of 1634 Adult Patients. *Epilepsy Behav.* **2004**, *5* (4), 593–595. https://doi.org/10.1016/j.yebeh.2004.03.009.

(39) Gerdung, C. A.; Tsang, A.; Yasseen, A. S.; Armstrong, K.; McMillan, H. J.; Kovesi, T. Association Between Chronic Aspiration and Chronic Airway Infection with Pseudomonas Aeruginosa and Other Gram-Negative Bacteria in Children with Cerebral Palsy. *Lung* **2016**, *194* (2), 307–314. https://doi.org/10.1007/s00408-016-9856-5.

(40) Restrepo, M. I.; Babu, B. L.; Reyes, L. F.; Chalmers, J. D.; Soni, N. J.; Sibila, O.; Faverio, P.; Cilloniz, C.; Rodriguez-Cintron, W.; Aliberti, S.; GLIMP. Burden and Risk Factors for Pseudomonas Aeruginosa Community-Acquired Pneumonia: A Multinational Point Prevalence Study of Hospitalised Patients. *Eur. Respir. J.* **2018**, *52* (2), 1701190. https://doi.org/10.1183/13993003.01190-2017.

(41) Santoro, S. L.; Chicoine, B.; Jasien, J. M.; Kim, J. L.; Stephens, M.; Bulova, P.; Capone, G. Pneumonia and Respiratory Infections in Down Syndrome: A Scoping Review of the Literature. *Am. J. Med. Genet. A.* **2021**, *185* (1), 286–299. https://doi.org/10.1002/ajmg.a.61924.

(42) McCaleb, R.; Warren, R. H.; Willis, D.; Maples, H. D.; Bai, S.; O'Brien, C. E. Description of Respiratory Microbiology of Children With Long-Term Tracheostomies. *Respir. Care* **2016**, *61* (4), 447–452. https://doi.org/10.4187/respcare.03518.
(43) Wilson, N. J.; Lin, Z.; Villarosa, A.; George, A. Oral Health Status and Reported Oral Health Problems in People with Intellectual Disability: A Literature Review. *J. Intellect. Dev. Disabil.* **2019**, *44* (3), 292–304. https://doi.org/10.3109/13668250.2017.1409596.
(44) Azarpazhooh, A.; Leake, J. L. Systematic Review of the Association Between Respiratory Diseases and Oral Health. *J. Periodontol.* **2006**, *77* (9), 1465–1482. https://doi.org/10.1902/jop.2006.060010.
(45) Son, M.; Jo, S.; Lee, J. S.; Lee, D. H. Association between Oral Health and Incidence of Pneumonia: A Population-Based Cohort Study from Korea. *Sci. Rep.* **2020**, *10* (1), 9576. https://doi.org/10.1038/s41598-020-66312-2.
(46) Emerson, E.; Robertson, J.; Baines, S.; Hatton, C. Obesity in British Children with and without Intellectual Disability: Cohort Study. *BMC Public Health* **2016**, *16* (1), 644. https://doi.org/10.1186/s12889-016-3309-1.
(47) Emerson, E. Underweight, Obesity and Exercise among Adults with Intellectual Disabilities in Supported Accommodation in Northern England. *J. Intellect. Disabil. Res. JIDR* **2005**, *49* (Pt 2), 134–143. https://doi.org/10.1111/j.1365-2788.2004.00617.x.
(48) Ranjan, S.; Nasser, J. A.; Fisher, K. Prevalence and Potential Factors Associated with Overweight and Obesity Status in Adults with Intellectual Developmental Disorders. *J. Appl. Res. Intellect. Disabil. JARID* **2018**, *31 Suppl 1*, 29–38. https://doi.org/10.1111/jar.12370.
(49) Phung, D. T.; Wang, Z.; Rutherford, S.; Huang, C.; Chu, C. Body Mass Index and Risk of Pneumonia: A Systematic Review and Meta-Analysis. *Obes. Rev. Off. J. Int. Assoc. Study Obes.* **2013**, *14* (10), 839–857. https://doi.org/10.1111/obr.12055.
(50) Anand, G.; Katz, P. O. Gastroesophageal Reflux Disease and Obesity. *Gastroenterol. Clin. North Am.* **2010**, *39* (1), 39–46. https://doi.org/10.1016/j.gtc.2009.12.002.
(51) Baskaran, V.; Murray, R. L.; Hunter, A.; Lim, W. S.; McKeever, T. M. Effect of Tobacco Smoking on the Risk of Developing Community Acquired Pneumonia: A Systematic Review and Meta-Analysis. *PLoS ONE* **2019**, *14* (7), e0220204. https://doi.org/10.1371/journal.pone.0220204.
(52) Vanker, A.; Gie, R. P.; Zar, H. J. The Association between Environmental Tobacco Smoke Exposure and Childhood Respiratory Disease: A Review. *Expert Rev. Respir. Med.* **2017**, *11* (8), 661–673. https://doi.org/10.1080/17476348.2017.1338949.
(53) Ahn, A.; Edwards, K. M.; Grijalva, C. G.; Self, W. H.; Zhu, Y.; Chappell, J. D.; Arnold, S. R.; McCullers, J. A.; Ampofo, K.; Pavia, A. T.; Bramley, A. M.; Jain, S.; Williams, D. J. Secondhand Smoke Exposure and Illness Severity Among Children Hospitalized with Pneumonia. *J. Pediatr.* **2015**, *167* (4), 869-874.e1. https://doi.org/10.1016/j.jpeds.2015.06.049.
(54) Torres, A.; Peetermans, W. E.; Viegi, G.; Blasi, F. Risk Factors for Community-Acquired Pneumonia in Adults in Europe: A Literature Review. *Thorax* **2013**, *68* (11), 1057–1065. https://doi.org/10.1136/thoraxjnl-2013-204282.
(55) Oulton, K. Personal Communication (In Press). **2021**.
(56) NHS. Reasonable adjustments https://www.england.nhs.uk/learning-disabilities/improving-health/reasonable-adjustments/.
(57) National Institute for Health and Care Excellence. Making Every Contact Count http://indepth.nice.org.uk/mecc/ (accessed 2021 -08 -23).
(58) Mencap. Communicating with people with a learning disability https://www.mencap.org.uk/learning-disability-explained/communicating-people-learning-disability (accessed 2021 -08 -23).
(59) Mencap. Health guides and Hospital Passports https://www.mencap.org.uk/advice-and-support/health/health-guides (accessed 2021 -09 -06).
(60) Office for Disability Issues, HM Government. Equality Act 2010: guidance https://www.gov.uk/guidance/equality-act-2010-guidance (accessed 2021 -08 -23).
(61) Leeds and York Partnership NHS Foundation Trust. Learning Disability Register Inclusion Tool. 2019.
(62) Carey, I. M.; Hosking, F. J.; Harris, T.; DeWilde, S.; Beighton, C.; Shah, S. M.; Cook, D. G. Do Health Checks for Adults with Intellectual Disability Reduce Emergency Hospital Admissions? Evaluation of a Natural Experiment. *J. Epidemiol. Community Health* **2016**, *71* (1), 52–58. https://doi.org/10.1136/jech-2016-207557.
(63) Heslop, P., Calkin, R., Byrne, V., Huxor, A., & Gielnik, K. Learning Disabilities Mortality Review (LeDeR) Programme Annual Report 2020. Bristol: Norah Fry Centre for Disability Studies, University of Bristol. 2021.
(64) NHS. Annual Health Checks.
(65) Slowie, D. and Martin, G. Narrowing the Health Inequality Gap by Annual Health Checks for Patients with Intellectual Disability. *BJGP* **2014**.
(66) BILD, Department of Health, & Mencap. Summary Report: Is Healthcare "Getting Better" for People with a Learning Disability? And Consultation Questionnaire Results: Is Healthcare "Getting Better" for People with a Learning Disability? 2013.

(67) Carey, I. M.; Shah, S. M.; Hosking, F. J.; DeWilde, S.; Harris, T.; Beighton, C.; Cook, D. G. Health Characteristics and Consultation Patterns of People with Intellectual Disability: A Cross-Sectional Database Study in English General Practice. *Br. J. Gen. Pract. J. R. Coll. Gen. Pract.* **2016**, *66* (645), e264-270. https://doi.org/10.3399/bjgp16X684301.
(68) Royal College of General Practitioners. Health Checks for People with Learning Disabilities Toolkit.
(69) Remedy: Bristol, North Somerset and South Gloucestershire Clinical Commissioning Group. Resources to Support Annual Health Checks for People with Learning Disabilities.
(70) Miller, M. R.; Hankinson, J.; Brusasco, V.; Burgos, F.; Casaburi, R.; Coates, A.; Crapo, R.; Enright, P.; van der Grinten, C. P. M.; Gustafsson, P.; Jensen, R.; Johnson, D. C.; MacIntyre, N.; McKay, R.; Navajas, D.; Pedersen, O. F.; Pellegrino, R.; Viegi, G.; Wanger, J.; ATS/ERS Task Force. Standardisation of Spirometry. *Eur. Respir. J.* **2005**, *26* (2), 319–338. https://doi.org/10.1183/09031936.05.00034805.
(71) Sylvester, K. P.; Clayton, N.; Cliff, I.; Hepple, M.; Kendrick, A.; Kirkby, J.; Miller, M.; Moore, A.; Rafferty, G. F.; O'Reilly, L.; Shakespeare, J.; Smith, L.; Watts, T.; Bucknall, M.; Butterfield, K. ARTP Statement on Pulmonary Function Testing 2020. *BMJ Open Respir. Res.* **2020**, *7* (1), e000575. https://doi.org/10.1136/bmjresp-2020-000575.
(72) Dohna-Schwake, C.; Ragette, R.; Teschler, H.; Voit, T.; Mellies, U. Predictors of Severe Chest Infections in Pediatric Neuromuscular Disorders. *Neuromuscul. Disord.* **2006**, *16* (5), 325–328. https://doi.org/10.1016/j.nmd.2006.02.003.
(73) Kulshrestha, R.; Willis, T.; Samuels, M. Noninvasive Ventilation in Paediatric Neuromuscular Disorders. In *Noninvasive Ventilation in Sleep Medicine and Pulmonary Critical Care: Critical Analysis of 2018-19 Clinical Trials*; Esquinas, A. M., Fiorentino, G., Insalaco, G., Mina, B., Duan, J., Mondardini, M. C., Caramelli, F., Eds.; Springer International Publishing: Cham, 2020; pp 505–514. https://doi.org/10.1007/978-3-030-42998-0_55.
(74) Fromageot, C.; Lofaso, F.; Annane, D.; Falaize, L.; Lejaille, M.; Clair, B.; Gajdos, P.; Raphaël, J. C. Supine Fall in Lung Volumes in the Assessment of Diaphragmatic Weakness in Neuromuscular Disorders. *Arch. Phys. Med. Rehabil.* **2001**, *82* (1), 123–128. https://doi.org/10.1053/apmr.2001.18053.
(75) Toussaint, M.; Chatwin, M.; Gonzales, J.; Berlowitz, D. J.; Enmc Respiratory Therapy Consortium. 228th ENMC International Workshop:: Airway Clearance Techniques in Neuromuscular Disorders Naarden, The Netherlands, 3-5 March, 2017. *Neuromuscul Disord* **2018**, *28* (3), 289–298. https://doi.org/10.1016/j.nmd.2017.10.008.
(76) Terzi, N.; Orlikowski, D.; Fermanian, C.; Lejaille, M.; Falaize, L.; Louis, A.; Raphael, J.-C.; Fauroux, B.; Lofaso, F. Measuring Inspiratory Muscle Strength in Neuromuscular Disease: One Test or Two? *Eur. Respir. J.* **2008**, *31* (1), 93–98. https://doi.org/10.1183/09031936.00094707.
(77) Fauroux, B.; Quijano-Roy, S.; Desguerre, I.; Khirani, S. The Value of Respiratory Muscle Testing in Children with Neuromuscular Disease. *Chest* **2015**, *147* (2), 552–559. https://doi.org/10.1378/chest.14-0819.
(78) Ragette, R.; Mellies, U.; Schwake, C.; Voit, T.; Teschler, H. Patterns and Predictors of Sleep Disordered Breathing in Primary Myopathies. *Thorax* **2002**, *57* (8), 724–728. https://doi.org/10.1136/thorax.57.8.724.
(79) Toussaint, M.; Steens, M.; Soudon, P. Lung Function Accurately Predicts Hypercapnia in Patients with Duchenne Muscular Dystrophy. *Chest* **2007**, *131* (2), 368–375. https://doi.org/10.1378/chest.06-1265.
(80) Stefanutti, D.; Benoist, M. R.; Scheinmann, P.; Chaussain, M.; Fitting, J. W. Usefulness of Sniff Nasal Pressure in Patients with Neuromuscular or Skeletal Disorders. *Am. J. Respir. Crit. Care Med.* **2000**, *162* (4 Pt 1), 1507–1511. https://doi.org/10.1164/ajrccm.162.4.9910034.
(81) Kaminska, M.; Noel, F.; Petrof, B. J. Optimal Method for Assessment of Respiratory Muscle Strength in Neuromuscular Disorders Using Sniff Nasal Inspiratory Pressure (SNIP). *PloS One* **2017**, *12* (5), e0177723. https://doi.org/10.1371/journal.pone.0177723.
(82) van de Wouw, E.; Evenhuis, H. M.; Echteld, M. A. Prevalence, Associated Factors and Treatment of Sleep Problems in Adults with Intellectual Disability: A Systematic Review. *Res. Dev. Disabil.* **2012**, *33* (4), 1310–1332. https://doi.org/10.1016/j.ridd.2012.03.003.
(83) Fan, Z.; Ahn, M.; Roth, H. L.; Li, L.; Vaughn, B. V. Sleep Apnea and Hypoventilation in Patients with Down Syndrome: Analysis of 144 Polysomnogram Studies. *Child. Basel* **2017**, *4* (7). https://doi.org/10.3390/children4070055.
(84) Blackmer, A. B.; Feinstein, J. A. Management of Sleep Disorders in Children With Neurodevelopmental Disorders: A Review. *Pharmacother. J. Hum. Pharmacol. Drug Ther.* **2016**, *36* (1), 84–98. https://doi.org/10.1002/phar.1686.
(85) Robinson-Shelton, A.; Malow, B. A. Sleep Disturbances in Neurodevelopmental Disorders. *Curr. Psychiatry Rep.* **2015**, *18* (1), 6. https://doi.org/10.1007/s11920-015-0638-1.
(86) Mellies, U.; Ragette, R.; Dohna Schwake, C.; Boehm, H.; Voit, T.; Teschler, H. Long-Term Noninvasive Ventilation in Children and Adolescents with Neuromuscular Disorders. *Eur. Respir. J.* **2003**, *22* (4), 631–636. https://doi.org/10.1183/09031936.03.00044303a.
(87) Toussaint, M.; Chatwin, M.; Soudon, P. Mechanical Ventilation in Duchenne Patients with Chronic Respiratory Insufficiency: Clinical Implications of 20 Years Published Experience. *Chron. Respir. Dis.* **2007**, *4* (3), 167–177. https://doi.org/10.1177/1479972307080697.

(88) Katz, S. L.; Gaboury, I.; Keilty, K.; Banwell, B.; Vajsar, J.; Anderson, P.; Ni, A.; Maclusky, I. Nocturnal Hypoventilation: Predictors and Outcomes in Childhood Progressive Neuromuscular Disease. *Arch. Dis. Child.* **2010**, *95* (12), 998–1003. https://doi.org/10.1136/adc.2010.182709.
(89) RCPCH. Working Party on Sleep Physiology and Respiratory Control Disorders in Childhood. Standards for Services for Children with Disorders of Sleep Physiology. RCPCH 2009.
(90) Gillett, E. S.; Perez, I. A. Disorders of Sleep and Ventilatory Control in Prader-Willi Syndrome. *Diseases* **2016**, *4* (3), 23. https://doi.org/10.3390/diseases4030023.
(91) Kapur, V. K.; Auckley, D. H.; Chowdhuri, S.; Kuhlmann, D. C.; Mehra, R.; Ramar, K.; Harrod, C. G. Clinical Practice Guideline for Diagnostic Testing for Adult Obstructive Sleep Apnea: An American Academy of Sleep Medicine Clinical Practice Guideline. *J. Clin. Sleep Med.* **2017**, *13* (03), 479–504. https://doi.org/10.5664/jcsm.6506.
(92) Evans, H, G., N. BTS Guideline for the Diagnosis and Monitoring of Paediatric Respiratory Sleep Disorders. Unpublished 2022.
(93) Zahm, J. M.; King, M.; Duvivier, C.; Pierrot, D.; Girod, S.; Puchelle, E. Role of Simulated Repetitive Coughing in Mucus Clearance. *Eur. Respir. J.* **1991**, *4* (3), 311–315.
(94) Leith, D. E. The Development of Cough. *Am. Rev. Respir. Dis.* **1985**, *131* (5), S39-42.
(95) Bianchi, C.; Baiardi, P. Cough Peak Flows: Standard Values for Children and Adolescents. *Am. J. Phys. Med. Rehabil.* **2008**, *87* (6), 461-467 10.1097/PHM.0b013e318174e4c7.
(96) Leith, D. E. Cough. In *Lung biology in health and disease:Respiratory defense mechanisms,. Cough*; New York: Marcel Deckker, 1977; pp 545–592.
(97) Szeinberg, A.; Tabachnik, E.; Rashed, N.; McLaughlin, F. J.; England, S.; Bryan, C. A.; Levison, H. Cough Capacity in Patients with Muscular Dystrophy. *Chest* **1988**, *94* (6), 1232–1235. https://doi.org/10.1378/chest.94.6.1232.
(98) Shneerson, J. M.; Simonds, A. K. Noninvasive Ventilation for Chest Wall and Neuromuscular Disorders. *Eur. Respir. J.* **2002**, *20* (2), 480–487. https://doi.org/10.1183/09031936.02.00404002.
(99) Galaz Souza, N.; Bush, A.; Tan, H.-L. Exploratory Study of the Effectiveness of Nebulised Saline in Children with Neurodisability. *Eur. Respir. J.* **2021**, *57* (3), 2001407. https://doi.org/10.1183/13993003.01407-2020.
(100) Chatwin, M.; Toussaint, M.; Goncalves, M. R.; Sheers, N.; Mellies, U.; Gonzales-Bermejo, J.; Sancho, J.; Fauroux, B.; Andersen, T.; Hov, B.; Nygren-Bonnier, M.; Lacombe, M.; Pernet, K.; Kampelmacher, M.; Devaux, C.; Kinnett, K.; Sheehan, D.; Rao, F.; Villanova, M.; Berlowitz, D.; Morrow, B. M. Airway Clearance Techniques in Neuromuscular Disorders: A State of the Art Review. *Respir Med* **2018**, *136*, 98–110. https://doi.org/10.1016/j.rmed.2018.01.012.
(101) Vitacca, M.; Paneroni, M.; Trainini, D.; Bianchi, L.; Assoni, G.; Saleri, M.; Gilè, S.; Winck, J. C.; Gonçalves, M. R. At Home and on Demand Mechanical Cough Assistance Program for Patients with Amyotrophic Lateral Sclerosis. *Am. J. Phys. Med. Rehabil.* **2010**, *89* (5), 401–406. https://doi.org/10.1097/PHM.0b013e3181d89760.
(102) WHO. WHO guidelines on physical activity and sedentary behaviour https://www.who.int/publications-detail-redirect/9789240015128 (accessed 2021 -09 -08).
(103) Dwyer, T. J.; Daviskas, E.; Zainuldin, R.; Verschuer, J.; Eberl, S.; Bye, P. T. P.; Alison, J. A. Effects of Exercise and Airway Clearance (Positive Expiratory Pressure) on Mucus Clearance in Cystic Fibrosis: A Randomised Crossover Trial. *Eur. Respir. J.* **2019**, *53* (4), 1801793. https://doi.org/10.1183/13993003.01793-2018.
(104) Dairo, Y. M.; Collett, J.; Dawes, H.; Oskrochi, G. R. Physical Activity Levels in Adults with Intellectual Disabilities: A Systematic Review. *Prev. Med. Rep.* **2016**, *4*, 209–219. https://doi.org/10.1016/j.pmedr.2016.06.008.
(105) Woodmansee, C.; Hahne, A.; Imms, C.; Shields, N. Comparing Participation in Physical Recreation Activities between Children with Disability and Children with Typical Development: A Secondary Analysis of Matched Data. *Res. Dev. Disabil.* **2016**, *49–50*, 268–276. https://doi.org/10.1016/j.ridd.2015.12.004.
(106) Shields, N.; Synnot, A. J.; Barr, M. Perceived Barriers and Facilitators to Physical Activity for Children with Disability: A Systematic Review. *Br. J. Sports Med.* **2012**, *46* (14), 989–997. https://doi.org/10.1136/bjsports-2011-090236.
(107) British Thoracic Society Standards of Care Committee. Non-Invasive Ventilation in Acute Respiratory Failure. *Thorax* **2002**, *57* (3), 192–211. https://doi.org/10.1136/thorax.57.3.192.
(108) Koontz, K. L.; Slifer, K. J.; Cataldo, M. D.; Marcus, C. L. Improving Pediatric Compliance with Positive Airway Pressure Therapy: The Impact of Behavioral Intervention. *Sleep* **2003**, *26* (8), 1010–1015. https://doi.org/10.1093/sleep/26.8.1010.
(109) Rapelli, G.; Pietrabissa, G.; Manzoni, G. M.; Bastoni, I.; Scarpina, F.; Tovaglieri, I.; Perger, E.; Garbarino, S.; Fanari, P.; Lombardi, C.; Castelnuovo, G. Improving CPAP Adherence in Adults With Obstructive Sleep Apnea Syndrome: A Scoping Review of Motivational Interventions. *Front. Psychol.* **2021**, *12*.
(110) Smith, P. L.; Wise, R. A.; Gold, A. R.; Schwartz, A. R.; Permutt, S. Upper Airway Pressure-Flow Relationships in Obstructive Sleep Apnea. *J. Appl. Physiol. Bethesda Md 1985* **1988**, *64* (2), 789–795. https://doi.org/10.1152/jappl.1988.64.2.789.

(111) Young, H. K.; Lowe, A.; Fitzgerald, D. A.; Seton, C.; Waters, K. A.; Kenny, E.; Hynan, L. S.; Iannaccone, S. T.; North, K. N.; Ryan, M. M. Outcome of Noninvasive Ventilation in Children with Neuromuscular Disease. *Neurology* **2007**, *68* (3), 198–201. https://doi.org/10.1212/01.wnl.0000251299.54608.13.

(112) Ward, S.; Chatwin, M.; Heather, S.; Simonds, A. K. Randomised Controlled Trial of Non-Invasive Ventilation (NIV) for Nocturnal Hypoventilation in Neuromuscular and Chest Wall Disease Patients with Daytime Normocapnia. *Thorax* **2005**, *60* (12), 1019–1024. https://doi.org/10.1136/thx.2004.037424.

(113) Dohna-Schwake, C.; Podlewski, P.; Voit, T.; Mellies, U. Non-Invasive Ventilation Reduces Respiratory Tract Infections in Children with Neuromuscular Disorders. *Pediatr. Pulmonol.* **2008**, *43* (1), 67–71. https://doi.org/10.1002/ppul.20740.

(114) Katz, S.; Selvadurai, H.; Keilty, K.; Mitchell, M.; MacLusky, I. Outcome of Non-Invasive Positive Pressure Ventilation in Paediatric Neuromuscular Disease. *Arch Child* **2004**, *89* (2), 121–124.

(115) Vianello, A.; Bevilacqua, M.; Salvador, V.; Cardaioli, C.; Vincenti, E. Long-Term Nasal Intermittent Positive Pressure Ventilation in Advanced Duchenne's Muscular Dystrophy. *Chest* **1994**, *105* (2), 445–448. https://doi.org/10.1378/chest.105.2.445.

(116) Oskoui, M.; Levy, G.; Garland, C. J.; Gray, J. M.; O'Hagen, J.; De Vivo, D. C.; Kaufmann, P. The Changing Natural History of Spinal Muscular Atrophy Type 1. *Neurology* **2007**, *69* (20), 1931–1936. https://doi.org/10.1212/01.wnl.0000290830.40544.b9.

(117) Chatwin, M.; Nickol, A. H.; Morrell, M. J.; Polkey, M. I.; Simonds, A. K. Randomised Trial of Inpatient versus Outpatient Initiation of Home Mechanical Ventilation in Patients with Nocturnal Hypoventilation. *Respir. Med.* **2008**, *102* (11), 1528–1535. https://doi.org/10.1016/j.rmed.2008.07.019.

(118) Pinto, T.; Chatwin, M.; Banfi, P.; Winck, J. C.; Nicolini, A. Mouthpiece Ventilation and Complementary Techniques in Patients with Neuromuscular Disease: A Brief Clinical Review and Update. *Chron. Respir. Dis.* **2017**, *14* (2), 187–193. https://doi.org/10.1177/1479972316674411.

(119) Fernandez-Bussy, S.; Mahajan, B.; Folch, E.; Caviedes, I.; Guerrero, J.; Majid, A. Tracheostomy Tube Placement: Early and Late Complications. *J. Bronchol. Interv. Pulmonol.* **2015**, *22* (4), 357–364. https://doi.org/10.1097/LBR.0000000000000177.

(120) Onakpoya, I. J.; Hayward, G.; Heneghan, C. J. The Use of Antibiotics for Preventing Lower Respiratory Tract Infections in at Risk Children Less than 12 Years Old. *Cochrane Database Syst. Rev.* **2015**, No. 2. https://doi.org/10.1002/14651858.CD011530.

(121) PARROT. Prophylactic Antibiotics to Prevent Chest Infections in Children with Neurological Impairment (PARROT) Trial. 2021.

(122) Hill, A. T.; Sullivan, A. L.; Chalmers, J. D.; De Soyza, A.; Elborn, S. J.; Floto, A. R.; Grillo, L.; Gruffydd-Jones, K.; Harvey, A.; Haworth, C. S.; Hiscocks, E.; Hurst, J. R.; Johnson, C.; Kelleher, P. W.; Bedi, P.; Payne, K.; Saleh, H.; Screaton, N. J.; Smith, M.; Tunney, M.; Whitters, D.; Wilson, R.; Loebinger, M. R. British Thoracic Society Guideline for Bronchiectasis in Adults. *Thorax* **2019**, *74* (Suppl 1), 1–69. https://doi.org/10.1136/thoraxjnl-2018-212463.

(123) Southern, K. W.; Barker, P. M.; Solis-Moya, A.; Patel, L. Macrolide Antibiotics for Cystic Fibrosis. *Cochrane Database Syst. Rev.* **2012**, *11*, CD002203. https://doi.org/10.1002/14651858.CD002203.pub4.

(124) Smith, D.; Du Rand, I.; Addy, C. L.; Collyns, T.; Hart, S. P.; Mitchelmore, P. J.; Rahman, N. M.; Saggu, R. British Thoracic Society Guideline for the Use of Long-Term Macrolides in Adults with Respiratory Disease. *Thorax* **2020**, *75* (5), 370–404. https://doi.org/10.1136/thoraxjnl-2019-213929.

(125) Garcia-Clemente, M.; de la Rosa, D.; Máiz, L.; Girón, R.; Blanco, M.; Olveira, C.; Canton, R.; Martinez-García, M. A. Impact of Pseudomonas Aeruginosa Infection on Patients with Chronic Inflammatory Airway Diseases. *J. Clin. Med.* **2020**, *9* (12). https://doi.org/10.3390/jcm9123800.

(126) van Ewijk, B. E.; Wolfs, T. F. W.; Fleer, A.; Kimpen, J. L. L.; van der Ent, C. K. High Pseudomonas Aeruginosa Acquisition Rate in CF. *Thorax* **2006**, *61* (7), 641–642. https://doi.org/10.1136/thx.2006.062372.

(127) Plioplys, A. V.; Kasnicka, I. Nebulized Tobramycin: Prevention of Pneumonias in Patients with Severe Cerebral Palsy. *J. Pediatr. Rehabil. Med.* **2011**, *4* (2), 155–158. https://doi.org/10.3233/PRM-2011-0168.

(128) Brodt, A. M.; Stovold, E.; Zhang, L. Inhaled Antibiotics for Stable Non-Cystic Fibrosis Bronchiectasis: A Systematic Review. *Eur. Respir. J.* **2014**, *44* (2), 382–393. https://doi.org/10.1183/09031936.00018414.

(129) Eckerland, M.; Bock, C.; Olivier, M.; Pichlmaier, L.; Steindor, M.; Stehling, F. Reducing the Frequency of Respiratory Tract Infections in Severe Neurological Disorders by Inhaled Antibiotics: A Retrospective Data Analysis. *ERJ Open Res.* **2019**, *5* (3), 00149–02018. https://doi.org/10.1183/23120541.00149-2018.

(130) Sanders, C. D.; Guimbellot, J. S.; Muhlebach, M. S.; Lin, F.-C.; Gilligan, P.; Esther, C. R. Tracheostomy in Children: Epidemiology and Clinical Outcomes. *Pediatr. Pulmonol.* **2018**, *53* (9), 1269–1275. https://doi.org/10.1002/ppul.24071.

(131) A John Simpson, Jamie-Leigh Allen, Michelle Chatwin, Hannah Crawford, Joanna Elverson, Victoria Ewan, Julian Forton, Ronan McMullan, John Plevris, Kate Renton, Hilary Tedd, Rhys Thomas,Julian Legg. BTS Clinical Statement: Aspiration Pneumonia. unpublished 2022.

(132) Logemann, J. A. *Evaluation and Treatment of Swallowing Disorders.*, 2nd ed.; 1999.

(133) van Timmeren, E. A.; van der Putten, A. a. J.; van Schrojenstein Lantman-de Valk, H. M. J.; van der Schans, C. P.; Waninge, A. Prevalence of Reported Physical Health Problems in People with Severe or Profound

Intellectual and Motor Disabilities: A Cross-Sectional Study of Medical Records and Care Plans. *J. Intellect. Disabil. Res. JIDR* **2016**, *60* (11), 1109–1118. https://doi.org/10.1111/jir.12298.

(134) Gal, E.; Hardal-Nasser, R.; Engel-Yeger, B. The Relationship between the Severity of Eating Problems and Intellectual Developmental Deficit Level. *Res. Dev. Disabil.* **2011**, *32* (5), 1464–1469. https://doi.org/10.1016/j.ridd.2010.12.003.

(135) Matson, J. L.; Fodstad, J. C.; Boisjoli, J. A. Cutoff Scores, Norms and Patterns of Feeding Problems for the Screening Tool of FEeding Problems (STEP) for Adults with Intellectual Disabilities. *Res. Dev. Disabil.* **2008**, *29* (4), 363–372. https://doi.org/10.1016/j.ridd.2007.06.001.

(136) Rogers, B. T.; Stratton, P.; Msall, M.; Andres, M.; Champlain, M. K.; Koerner, P.; Piazza, J. Long-Term Morbidity and Management Strategies of Tracheal Aspiration in Adults with Severe Developmental Disabilities. *Am. J. Ment. Retard.* **1994**, *98* (4), 490–498.

(137) Hickman, J.; Jenner, J. ALD and Dysphagia: Issues and Practice. *Speech Lang Ther Pr.* **1997**, 8–11.

(138) Thacker, A.; Abdelnoor, A.; Anderson, C.; White, S.; Hollins, S. Indicators of Choking Risk in Adults with Learning Disabilities: A Questionnaire Survey and Interview Study. *Disabil. Rehabil.* **2008**, *30* (15), 1131–1138. https://doi.org/10.1080/09638280701461625.

(139) Aziz, S. J.; Campbell-Taylor, I. Neglect and Abuse Associated with Undernutrition in Long-Term Care in North America: Causes and Solutions. *J. Elder Abuse Negl.* **1999**, *10* (1–2), 91–117. https://doi.org/10.1300/J084v10n01_07.

(140) Eyman, R. K.; Grossman, H. J.; Chaney, R. H.; Call, T. L. The Life Expectancy of Profoundly Handicapped People with Mental Retardation. *N. Engl. J. Med.* **1990**, *323* (9), 584–589. https://doi.org/10.1056/NEJM199008303230906.

(141) Beange, H.; McElduff, A.; Baker, W. Medical Disorders of Adults with Mental Retardation: A Population Study. *Am. J. Ment. Retard. AJMR* **1995**, *99* (6), 595–604.

(142) Jacobsson, C.; Axelsson, K.; Osterlind, P. O.; Norberg, A. How People with Stroke and Healthy Older People Experience the Eating Process. *J. Clin. Nurs.* **2000**, *9* (2), 255–264. https://doi.org/10.1046/j.1365-2702.2000.00355.x.

(143) National Patient Safety Agency. Understanding the Patient Safety Issues for People with Learning Disabilities. 2004.

(144) Gallagher, L.; Naidoo, P. Prescription Drugs and Their Effects on Swallowing. *Dysphagia* **2009**, *24* (2), 159–166. https://doi.org/10.1007/s00455-008-9187-7.

(145) Manduchi, B.; Fainman, G. M.; Walshe, M. Interventions for Feeding and Swallowing Disorders in Adults with Intellectual Disability: A Systematic Review of the Evidence. *Dysphagia* **2020**, *35* (2), 207–219. https://doi.org/10.1007/s00455-019-10038-5.

(146) Sheehan, R.; Hassiotis, A.; Walters, K.; Osborn, D.; Strydom, A.; Horsfall, L. Mental Illness, Challenging Behaviour, and Psychotropic Drug Prescribing in People with Intellectual Disability: UK Population Based Cohort Study. *BMJ* **2015**, *351*, h4326. https://doi.org/10.1136/bmj.h4326.

(147) Rumbach, A.; Coombes, C.; Doeltgen, S. A Survey of Australian Dysphagia Practice Patterns. *Dysphagia* **2018**, *33* (2), 216–226. https://doi.org/10.1007/s00455-017-9849-4.

(148) Hendrix, T. R. Art and Science of History Taking in the Patient with Difficulty Swallowing. *Dysphagia* **1993**, *8* (2), 69–73. https://doi.org/10.1007/BF02266982.

(149) Speyer, R. Oropharyngeal Dysphagia: Screening and Assessment. *Otolaryngol. Clin. North Am.* **2013**, *46* (6), 989–1008. https://doi.org/10.1016/j.otc.2013.08.004.

(150) McAllister, S.; Kruger, S.; Doeltgen, S.; Tyler-Boltrek, E. Implications of Variability in Clinical Bedside Swallowing Assessment Practices by Speech Language Pathologists. *Dysphagia* **2016**, *31* (5), 650–662. https://doi.org/10.1007/s00455-016-9724-8.

(151) Bateman, C.; Leslie, P.; Drinnan, M. J. Adult Dysphagia Assessment in the UK and Ireland: Are SLTs Assessing the Same Factors? *Dysphagia* **2007**, *22* (3), 174–186. https://doi.org/10.1007/s00455-006-9070-3.

(152) Martino, R.; Pron, G.; Diamant, N. E. Oropharyngeal Dysphagia: Surveying Practice Patterns of the Speech-Language Pathologist. *Dysphagia* **2004**, *19* (3), 165–176. https://doi.org/10.1007/s00455-004-0004-7.

(153) Vogels, B.; Cartwright, J.; Cocks, N. The Bedside Assessment Practices of Speech-Language Pathologists in Adult Dysphagia. *Int. J. Speech Lang. Pathol.* **2015**, *17* (4), 390–400. https://doi.org/10.3109/17549507.2014.979877.

(154) Hungin, A. P. S.; Molloy-Bland, M.; Scarpignato, C. Revisiting Montreal: New Insights into Symptoms and Their Causes, and Implications for the Future of GERD. *Am. J. Gastroenterol.* **2019**, *114* (3), 414–421. https://doi.org/10.1038/s41395-018-0287-1.

(155) Morehead, R. S. Gastro-Oesophageal Reflux Disease and Non-Asthma Lung Disease. *Eur. Respir. Rev. Off. J. Eur. Respir. Soc.* **2009**, *18* (114), 233–243. https://doi.org/10.1183/09059180.00002509.

(156) Lee, A. S.; Lee, J. S.; He, Z.; Ryu, J. H. Reflux-Aspiration in Chronic Lung Disease. *Ann. Am. Thorac. Soc.* **2020**, *17* (2), 155–164. https://doi.org/10.1513/AnnalsATS.201906-427CME.

(157) Bozkurt, M.; Tutuncuoglu, S.; Serdaroglu, G.; Tekgul, H.; Aydogdu, S. Gastroesophageal Reflux in Children with Cerebral Palsy: Efficacy of Cisapride. *J. Child Neurol.* **2004**, *19* (12), 973–976.

(158) Gangil, A.; Patwari, A. K.; Bajaj, P.; Kashyap, R.; Anand, V. K. Gastroesophageal Reflux Disease in Children with Cerebral Palsy. *Indian Pediatr.* **2001**, *38* (7), 766–770.

(159) Reyes, A. L.; Cash, A. J.; Green, S. H.; Booth, I. W. Gastrooesophageal Reflux in Children with Cerebral Palsy. *Child Care Health Dev.* **1993**, *19* (2), 109–118. https://doi.org/10.1111/j.1365-2214.1993.tb00718.x.

(160) de Veer, A. J.; Bos, J. T.; Boer, R. C. N.; Böhmer, C. J.; Francke, A. L. Symptoms of Gastroesophageal Reflux Disease in Severely Mentally Retarded People: A Systematic Review. *BMC Gastroenterol.* **2008**, *8*, 23. https://doi.org/10.1186/1471-230X-8-23.

(161) Romano, C.; van Wynckel, M.; Hulst, J.; Broekaert, I.; Bronsky, J.; Dall'Oglio, L.; Mis, N. F.; Hojsak, I.; Orel, R.; Papadopoulou, A.; Schaeppi, M.; Thapar, N.; Wilschanski, M.; Sullivan, P.; Gottrand, F. European Society for Paediatric Gastroenterology, Hepatology and Nutrition Guidelines for the Evaluation and Treatment of Gastrointestinal and Nutritional Complications in Children With Neurological Impairment. *J. Pediatr. Gastroenterol. Nutr.* **2017**, *65* (2), 242–264. https://doi.org/10.1097/MPG.0000000000001646.

(162) Ang, D.; Lee, Y.-Y.; Clarke, J. O.; Lynch, K.; Guillaume, A.; Onyimba, F.; Kamal, A.; Gyawali, C. P. Diagnosis of Gastroesophageal Reflux: An Update on Current and Emerging Modalities. *Ann. N. Y. Acad. Sci.* **2020**, *1481* (1), 154–169. https://doi.org/10.1111/nyas.14369.

(163) Rosen, R.; Vandenplas, Y.; Singendonk, M.; Cabana, M.; Di Lorenzo, C.; Gottrand, F.; Gupta, S.; Langendam, M.; Staiano, A.; Thapar, N.; Tipnis, N.; Tabbers, M. Pediatric Gastroesophageal Reflux Clinical Practice Guidelines: Joint Recommendations of the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) and the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition (ESPGHAN). *J. Pediatr. Gastroenterol. Nutr.* **2018**, *66* (3), 516–554. https://doi.org/10.1097/MPG.0000000000001889.

(164) Katz, P. O.; Gerson, L. B.; Vela, M. F. Guidelines for the Diagnosis and Management of Gastroesophageal Reflux Disease. *Off. J. Am. Coll. Gastroenterol. ACG* **2013**, *108* (3), 308–328. https://doi.org/10.1038/ajg.2012.444.

(165) Wang, C.-H.; Li, C.-H.; Hsieh, R.; Fan, C.-Y.; Hsu, T.-C.; Chang, W.-C.; Hsu, W.-T.; Lin, Y.-Y.; Lee, C.-C. Proton Pump Inhibitors Therapy and the Risk of Pneumonia: A Systematic Review and Meta-analysis of Randomized Controlled Trials and Observational Studies. *Expert Opin. Drug Saf.* **2019**, *18* (3), 163–172. https://doi.org/10.1080/14740338.2019.1577820.

(166) Fuchs, H. F.; Gutschow, C. A.; Brinkmann, S.; Herbold, T.; Bludau, M.; Schröder, W.; Bollschweiler, E.; Hölscher, A. H.; Leers, J. M. Effect of Laparoscopic Antireflux Surgery on Esophageal Motility. *Dig. Surg.* **2014**, *31* (4–5), 354–358. https://doi.org/10.1159/000368662.

(167) Mauritz, F. A.; van Herwaarden-Lindeboom, M. Y. A.; Stomp, W.; Zwaveling, S.; Fischer, K.; Houwen, R. H. J.; Siersema, P. D.; van der Zee, D. C. The Effects and Efficacy of Antireflux Surgery in Children with Gastroesophageal Reflux Disease: A Systematic Review. *J. Gastrointest. Surg. Off. J. Soc. Surg. Aliment. Tract* **2011**, *15* (10), 1872–1878. https://doi.org/10.1007/s11605-011-1644-1.

(168) Rickenbacher, N.; Kötter, T.; Kochen, M. M.; Scherer, M.; Blozik, E. Fundoplication versus Medical Management of Gastroesophageal Reflux Disease: Systematic Review and Meta-Analysis. *Surg. Endosc.* **2014**, *28* (1), 143–155. https://doi.org/10.1007/s00464-013-3140-z.

(169) Maret-Ouda, J.; Santoni, G.; Artama, M.; Ness-Jensen, E.; Svensson, J. F.; von Euler-Chelpin, M.; Lagergren, J. Aspiration Pneumonia after Antireflux Surgery among Neurologically Impaired Children with GERD. *J. Pediatr. Surg.* **2020**, *55* (11), 2408–2412. https://doi.org/10.1016/j.jpedsurg.2019.12.024.

(170) Broekaert, I. J.; Falconer, J.; Bronsky, J.; Gottrand, F.; Dall'Oglio, L.; Goto, E.; Hojsak, I.; Hulst, J.; Kochavi, B.; Papadopoulou, A.; Ribes-Koninckx, C.; Schaeppi, M.; Werlin, S.; Wilschanski, M.; Thapar, N. The Use of Jejunal Tube Feeding in Children: A Position Paper by the Gastroenterology and Nutrition Committees of the European Society for Paediatric Gastroenterology, Hepatology, and Nutrition 2019. *J. Pediatr. Gastroenterol. Nutr.* **2019**, *69* (2), 239–258. https://doi.org/10.1097/MPG.0000000000002379.

(171) Livingston, M. H.; Shawyer, A. C.; Rosenbaum, P. L.; Jones, S. A.; Walton, J. M. Fundoplication and Gastrostomy versus Percutaneous Gastrojejunostomy for Gastroesophageal Reflux in Children with Neurologic Impairment: A Systematic Review and Meta-Analysis. *J. Pediatr. Surg.* **2015**, *50* (5), 707–714. https://doi.org/10.1016/j.jpedsurg.2015.02.020.

(172) Srivastava, R.; Downey, E. C.; O'Gorman, M.; Feola, P.; Samore, M.; Holubkov, R.; Mundorff, M.; James, B. C.; Rosenbaum, P.; Young, P. C.; Dean, J. M. Impact of Fundoplication versus Gastrojejunal Feeding Tubes on Mortality and in Preventing Aspiration Pneumonia in Young Children with Neurologic Impairment Who Have Gastroesophageal Reflux Disease. *Pediatrics* **2009**, *123* (1), 338–345. https://doi.org/10.1542/peds.2007-1740.

(173) Rashnoo, P.; Daniel, S. J. Drooling Quantification: Correlation of Different Techniques. *Int. J. Pediatr. Otorhinolaryngol.* **2015**, *79* (8), 1201–1205. https://doi.org/10.1016/j.ijporl.2015.05.010.

(174) Fairhurst, C. B. R.; Cockerill, H. Management of Drooling in Children. *Arch. Dis. Child. Educ. Pract. Ed.* **2011**, *96* (1), 25–30. https://doi.org/10.1136/adc.2007.129478.

(175) Varley, L. P.; Denieffe, S.; O'Gorman, C.; Murphy, A.; Gooney, M. A Systematic Review of Noninvasive and Invasive Sialorrhoea Management. *J. Clin. Nurs.* **2019**, *28* (23–24), 4190–4206. https://doi.org/10.1111/jocn.15009.

(176) American Academy for Cerebral Palsy and Developmental Medicine. Care Pathways: Sialorrhea in Cerebral Palsy. 2016.

(177) Parr, J. R.; Todhunter, E.; Pennington, L.; Stocken, D.; Cadwgan, J.; O'Hare, A. E.; Tuffrey, C.; Williams, J.; Cole, M.; Colver, A. F. Drooling Reduction Intervention Randomised Trial (DRI): Comparing the Efficacy and Acceptability of Hyoscine Patches and Glycopyrronium Liquid on Drooling in Children with Neurodisability. *Arch. Dis. Child.* **2018**, *103* (4), 371–376. https://doi.org/10.1136/archdischild-2017-313763.

(178) National Institute for Health and Care Excellence. Severe sialorrhoea (drooling) in children and young people with chronic neurological disorders: oral glycopyrronium bromide https://www.nice.org.uk/advice/es5/chapter/Key-points (accessed 2021 -08 -17).

(179) Norderyd, J.; Graf, J.; Marcusson, A.; Nilsson, K.; Sjöstrand, E.; Steinwall, G.; Ärleskog, E.; Bågesund, M. Sublingual Administration of Atropine Eyedrops in Children with Excessive Drooling – a Pilot Study. *Int. J. Paediatr. Dent.* **2017**, *27* (1), 22–29. https://doi.org/10.1111/ipd.12219.

(180) Jost, W. H.; Friedman, A.; Michel, O.; Oehlwein, C.; Slawek, J.; Bogucki, A.; Ochudlo, S.; Banach, M.; Pagan, F.; Flatau-Baqué, B.; Csikós, J.; Cairney, C. J.; Blitzer, A. SIAXI: Placebo-Controlled, Randomized, Double-Blind Study of IncobotulinumtoxinA for Sialorrhea. *Neurology* **2019**, *92* (17), e1982–e1991. https://doi.org/10.1212/WNL.0000000000007368.

(181) National Institute for Health and Care Excellence. Xeomin (botulinum neurotoxin type A) for treating chronic sialorrhoea https://www.nice.org.uk/guidance/ta605 (accessed 2021 -08 -17).

(182) Dohar, J. E. Sialorrhea & Aspiration Control - A Minimally Invasive Strategy Uncomplicated by Anticholinergic Drug Tolerance or Tachyphylaxis. *Int. J. Pediatr. Otorhinolaryngol.* **2019**, *116*, 97–101. https://doi.org/10.1016/j.ijporl.2018.10.035.

(183) 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* (12), 2124–2128. https://doi.org/10.1016/j.ijporl.2015.09.029.

(184) Lawrence, R.; Bateman, N. Surgical Management of the Drooling Child. *Curr. Otorhinolaryngol. Rep.* **2018**, *6* (1), 99–106. https://doi.org/10.1007/s40136-018-0188-2.

(185) Gerber, M. E.; Gaugler, M. D.; Myer, C. M.; Cotton, R. T. Chronic Aspiration in Children. When Are Bilateral Submandibular Gland Excision and Parotid Duct Ligation Indicated? *Arch. Otolaryngol. Head Neck Surg.* **1996**, *122* (12), 1368–1371. https://doi.org/10.1001/archotol.1996.01890240074016.

(186) Manrique, D.; Sato, J. Salivary Gland Surgery for Control of Chronic Pulmonary Aspiration in Children with Cerebral Palsy. *Int. J. Pediatr. Otorhinolaryngol.* **2009**, *73* (9), 1192–1194. https://doi.org/10.1016/j.ijporl.2009.05.002.

(187) Robertson, J.; Hatton, C.; Emerson, E.; Baines, S. Prevalence of Epilepsy among People with Intellectual Disabilities: A Systematic Review. *Seizure* **2015**, *29*, 46–62. https://doi.org/10.1016/j.seizure.2015.03.016.

(188) National Institute for Health and Care Excellence. Epilepsies: diagnosis and management (CG137) https://www.nice.org.uk/guidance/cg137 (accessed 2021 -09 -06).

(189) Gomes-Filho, I. S.; Passos, J. S.; Seixas da Cruz, S. Respiratory Disease and the Role of Oral Bacteria. *J. Oral Microbiol.* **2010**, *2*. https://doi.org/10.3402/jom.v2i0.5811.

(190) Khadka, S.; Khan, S.; King, A.; Goldberg, L. R.; Crocombe, L.; Bettiol, S. Poor Oral Hygiene, Oral Microorganisms and Aspiration Pneumonia Risk in Older People in Residential Aged Care: A Systematic Review. *Age Ageing* **2021**, *50* (1), 81–87. https://doi.org/10.1093/ageing/afaa102.

(191) Manger, D.; Walshaw, M.; Fitzgerald, R.; Doughty, J.; Wanyonyi, K. L.; White, S.; Gallagher, J. E. Evidence Summary: The Relationship between Oral Health and Pulmonary Disease. *Br. Dent. J.* **2017**, *222* (7), 527–533. https://doi.org/10.1038/sj.bdj.2017.315.

(192) Sjögren, P.; Nilsson, E.; Forsell, M.; Johansson, O.; Hoogstraate, J. A Systematic Review of the Preventive Effect of Oral Hygiene on Pneumonia and Respiratory Tract Infection in Elderly People in Hospitals and Nursing Homes: Effect Estimates and Methodological Quality of Randomized Controlled Trials. *J. Am. Geriatr. Soc.* **2008**, *56* (11), 2124–2130. https://doi.org/10.1111/j.1532-5415.2008.01926.x.

(193) Lin, J. L.; Van Haren, K.; Rigdon, J.; Saynina, O.; Song, H.; Buu, M. C.; Thakur, Y.; Srinivas, N.; Asch, S. M.; Sanders, L. M. Pneumonia Prevention Strategies for Children With Neurologic Impairment. *Pediatrics* **2019**, *144* (4). https://doi.org/10.1542/peds.2019-0543.

(194) Public Health England. Oral care and people with learning disabilities https://www.gov.uk/government/publications/oral-care-and-people-with-learning-disabilities/oral-care-and-people-with-learning-disabilities (accessed 2021 -08 -21).

(195) Nie, W.; Zhang, Y.; Jee, S. H.; Jung, K. J.; Li, B.; Xiu, Q. Obesity Survival Paradox in Pneumonia: A Meta-Analysis. *BMC Med.* **2014**, *12* (1), 61. https://doi.org/10.1186/1741-7015-12-61.

(196) National Institute for Health and Care Excellence. Overview | Nutrition support for adults: oral nutrition support, enteral tube feeding and parenteral nutrition | Guidance | NICE https://www.nice.org.uk/guidance/cg32 (accessed 2021 -08 -18).

(197) National Institute for Health and Care Excellence. Weight management: lifestyle services for overweight or obese children and young people https://www.nice.org.uk/guidance/ph47 (accessed 2021 -08 -18).

(198) National Institute for Health and Care Excellence. Obesity: identification, assessment and management https://www.nice.org.uk/guidance/cg189 (accessed 2021 -08 -18).

(199) National Institute for Health and Care Excellence. Faltering growth: recognition and management of faltering growth in children (NG75) https://www.nice.org.uk/guidance/ng75/chapter/Recommendations#faltering-growth-after-the-early-days-of-life (accessed 2021 -12 -05).
(200) Samson-Fang, L. J.; Stevenson, R. D. Identification of Malnutrition in Children with Cerebral Palsy: Poor Performance of Weight-for-Height Centiles. *Dev. Med. Child Neurol.* **2000**, *42* (3), 162–168. https://doi.org/10.1017/s0012162200000293.
(201) Kuperminc, M. N.; Gurka, M. J.; Bennis, J. A.; Busby, M. G.; Grossberg, R. I.; Henderson, R. C.; Stevenson, R. D. Anthropometric Measures: Poor Predictors of Body Fat in Children with Moderate to Severe Cerebral Palsy. *Dev. Med. Child Neurol.* **2010**, *52* (9), 824–830. https://doi.org/10.1111/j.1469-8749.2010.03694.x.
(202) Spanos, D.; Hankey, C.; Boyle, S.; Melville, C. Comparing the Effectiveness of a Multi-Component Weight Loss Intervention in Adults with and without Intellectual Disabilities. *J. Hum. Nutr. Diet. Off. J. Br. Diet. Assoc.* **2014**, *27* (1), 22–29. https://doi.org/10.1111/jhn.12051.
(203) Heo, J. Y.; Song, J. Y.; Noh, J. Y.; Choi, M. J.; Yoon, J. G.; Lee, S. N.; Cheong, H. J.; Kim, W. J. Effects of Influenza Immunization on Pneumonia in the Elderly. *Hum. Vaccines Immunother.* **2018**, *14* (3), 744–749. https://doi.org/10.1080/21645515.2017.1405200.
(204) van Werkhoven, C. H.; Huijts, S. M. Vaccines to Prevent Pneumococcal Community-Acquired Pneumonia. *Clin. Chest Med.* **2018**, *39* (4), 733–752. https://doi.org/10.1016/j.ccm.2018.07.007.
(205) Fiore, A. E.; Bridges, C. B.; Cox, N. J. Seasonal Influenza Vaccines. In *Vaccines for Pandemic Influenza*; Compans, R. W., Orenstein, W. A., Eds.; Current Topics in Microbiology and Immunology; Springer: Berlin, Heidelberg, 2009; pp 43–82. https://doi.org/10.1007/978-3-540-92165-3_3.
(206) Emerson, E.; Robertson, J.; Baines, S.; Hatton, C. Vaccine Coverage among Children with and without Intellectual Disabilities in the UK: Cross Sectional Study. *BMC Public Health* **2019**, *19*, 748. https://doi.org/10.1186/s12889-019-7106-5.
(207) Weise, J. C.; Srasuebkul, P.; Trollor, J. N. Potentially Preventable Hospitalisations of People with Intellectual Disability in New South Wales. *Med. J. Aust.* **2021**, *215* (1).
(208) Public Health England. Flu vaccinations: supporting people with learning disabilities https://www.gov.uk/government/publications/flu-vaccinations-for-people-with-learning-disabilities/flu-vaccinations-supporting-people-with-learning-disabilities (accessed 2021 -08 -17).
(209) Public Health England. Information on how to protect yourself from flu: easy-read guide https://www.gov.uk/government/publications/flu-leaflet-for-people-with-learning-disability/information-on-how-to-protect-yourself-from-flu-easy-read (accessed 2021 -08 -17).
(210) Pneumococcal vaccine and Learning disability - a Freedom of Information request to The Joint Committee on Vaccination and Immunisation https://www.whatdotheyknow.com/request/pneumococcal_vaccine_and_learnin (accessed 2021 -08 -17).
(211) Public Health England. Immunisation against Infectious Disease.
(212) Public Health England. Pneumococcal: the green book, chapter 25 https://www.gov.uk/government/publications/pneumococcal-the-green-book-chapter-25 (accessed 2021 -08 -17).
(213) Public Health England. Consent: the green book, chapter 2 https://www.gov.uk/government/publications/consent-the-green-book-chapter-2 (accessed 2021 -08 -17).
(214) *Mental Capacity Act 2005: Code of Practice*, 2007 final ed.; Great Britain, Ed.; TSO: London, 2007.
(215) Northern Ireland Assembly. Mental Capacity Act (Northern Ireland) 2016 https://www.legislation.gov.uk/nia/2016/18/contents/enacted (accessed 2021 -12 -01).
(216) Scottish Parliament. Adults with Incapacity (Scotland) Act 2000 https://www.legislation.gov.uk/asp/2000/4/contents (accessed 2021 -12 -01).
(217) British Thoracic Society. Annotated BTS Guideline for the Management of CAP in Adults. 2015.
(218) Ksinan, A. J.; Sheng, Y.; Do, E. K.; Schechter, J. C.; Zhang, J. (Jim); Maguire, R. L.; Hoyo, C.; Murphy, S. K.; Kollins, S. H.; Rubin, B.; Fuemmeler, B. F. Identifying the Best Questions for Rapid Screening of Secondhand Smoke Exposure Among Children. *Nicotine Tob. Res.* **2021**, *23* (7), 1217–1223. https://doi.org/10.1093/ntr/ntaa254.
(219) Stop Smoking Interventions and Services. NG92. National Institute for Health and Care Excellence 2018.
(220) Harris, M.; Clark, J.; Coote, N.; Fletcher, P.; Harnden, A.; McKean, M.; Thomson, A. British Thoracic Society Guidelines for the Management of Community Acquired Pneumonia in Children: Update 2011. *Thorax* **2011**, *66* (Suppl 2), ii1–ii23. https://doi.org/10.1136/thoraxjnl-2011-200598.
(221) National Institute for Health and Care Excellence. Pneumonia (community-acquired): antimicrobial prescribing https://www.nice.org.uk/guidance/ng138/chapter/recommendations (accessed 2021 -08 -18).
(222) Rudan, I.; Boschi-Pinto, C.; Biloglav, Z.; Mulholland, K.; Campbell, H. Epidemiology and Etiology of Childhood Pneumonia. *Bull. World Health Organ.* **2008**, *86* (5), 408–416. https://doi.org/10.2471/blt.07.048769.
(223) Binkley, C. J.; Haugh, G. S.; Kitchens, D. H.; Wallace, D. L.; Sessler, D. I. Oral Microbial and Respiratory Status of Persons with Mental Retardation/Intellectual and Developmental Disability: An Observational

Cohort Study. *Oral Surg. Oral Med. Oral Pathol. Oral Radiol. Endod.* **2009**, *108* (5), 722–731. https://doi.org/10.1016/j.tripleo.2009.06.027.

(224) Ieven, M.; Coenen, S.; Loens, K.; Lammens, C.; Coenjaerts, F.; Vanderstraeten, A.; Henriques-Normark, B.; Crook, D.; Huygen, K.; Butler, C. C.; Verheij, T. J. M.; Little, P.; Zlateva, K.; van Loon, A.; Claas, E. C. J.; Goossens, H. Aetiology of Lower Respiratory Tract Infection in Adults in Primary Care: A Prospective Study in 11 European Countries. *Clin. Microbiol. Infect.* **2018**, *24* (11), 1158–1163. https://doi.org/10.1016/j.cmi.2018.02.004.

(225) Bhuiyan, M. U.; Snelling, T. L.; West, R.; Lang, J.; Rahman, T.; Granland, C.; Gier, C. de; Borland, M. L.; Thornton, R. B.; Kirkham, L.-A. S.; Sikazwe, C.; Martin, A. C.; Richmond, P. C.; Smith, D. W.; Jaffe, A.; Blyth, C. C. The Contribution of Viruses and Bacteria to Community-Acquired Pneumonia in Vaccinated Children: A Case–Control Study. *Thorax* **2019**, *74* (3), 261–269. https://doi.org/10.1136/thoraxjnl-2018-212096.

(226) Jain, S.; Williams, D. J.; Arnold, S. R.; Ampofo, K.; Bramley, A. M.; Reed, C.; Stockmann, C.; Anderson, E. J.; Grijalva, C. G.; Self, W. H.; Zhu, Y.; Patel, A.; Hymas, W.; Chappell, J. D.; Kaufman, R. A.; Kan, J. H.; Dansie, D.; Lenny, N.; Hillyard, D. R.; Haynes, L. M.; Levine, M.; Lindstrom, S.; Winchell, J. M.; Katz, J. M.; Erdman, D.; Schneider, E.; Hicks, L. A.; Wunderink, R. G.; Edwards, K. M.; Pavia, A. T.; McCullers, J. A.; Finelli, L.; CDC EPIC Study Team. Community-Acquired Pneumonia Requiring Hospitalization among U.S. Children. *N. Engl. J. Med.* **2015**, *372* (9), 835–845. https://doi.org/10.1056/NEJMoa1405870.

(227) Meskill, S. D.; O'Bryant, S. C. Respiratory Virus Co-Infection in Acute Respiratory Infections in Children. *Curr. Infect. Dis. Rep.* **2020**, *22* (1), 3. https://doi.org/10.1007/s11908-020-0711-8.

(228) Hampshire, Southampton and Isle of Wight CCG. RESTORE2 https://www.hampshiresouthamptonandisleofwightccg.nhs.uk/your-health/restore-official (accessed 2022 -02 -08).

(229) Shah, S. N. M.; Bachur, R. G.; Simel, D. L.; Neuman, M. I. M. Does This Child Have Pneumonia?: The Rational Clinical Examination Systematic Review. [Review]. *JAMA* **2017**, *318* (5), 462–471. https://doi.org/10.1001/jama.2017.9039.

(230) Koumbourlis, A. C. Scoliosis and the Respiratory System. *Paediatr. Respir. Rev.* **2006**, *7* (2), 152–160. https://doi.org/10.1016/j.prrv.2006.04.009.

(231) Lim, W. S.; van der Eerden, M. M.; Laing, R.; Boersma, W. G.; Karalus, N.; Town, G. I.; Lewis, S. A.; Macfarlane, J. T. Defining Community Acquired Pneumonia Severity on Presentation to Hospital: An International Derivation and Validation Study. *Thorax* **2003**, *58* (5), 377–382. https://doi.org/10.1136/thorax.58.5.377.

(232) Hanson, K. E.; Azar, M. M.; Banerjee, R.; Chou, A.; Colgrove, R. C.; Ginocchio, C. C.; Hayden, M. K.; Holodiny, M.; Jain, S.; Koo, S.; Levy, J.; Timbrook, T. T.; Caliendo, A. M.; for the Diagnostics Committee of the Infectious Diseases Society of America (IDSA). Molecular Testing for Acute Respiratory Tract Infections: Clinical and Diagnostic Recommendations From the IDSA's Diagnostics Committee. *Clin. Infect. Dis.* **2020**, *71* (10), 2744–2751. https://doi.org/10.1093/cid/ciaa508.

(233) British HIV association. BHIVA/BASHH/BIA Adult HIV Testing guidelines 2020 https://www.bhiva.org/HIV-testing-guidelines (accessed 2021 -08 -18).

(234) Silverston, P. Pulse Oximetry in Primary Care. *InnovAiT* **2016**, *9* (4), 202–207. https://doi.org/10.1177/1755738016628894.

(235) Principi, N.; Esposito, S. Biomarkers in Pediatric Community-Acquired Pneumonia. *Int. J. Mol. Sci.* **2017**, *18* (2). https://doi.org/10.3390/ijms18020447.

(236) Royal College of Physicians. National Early Warning Score (NEWS) 2 https://www.rcplondon.ac.uk/projects/outputs/national-early-warning-score-news-2 (accessed 2021 -08 -18).

(237) O'Driscoll, B. R.; Howard, L. S.; Earis, J.; Mak, V.; British Thoracic Society Emergency Oxygen Guideline Group; BTS Emergency Oxygen Guideline Development Group. BTS Guideline for Oxygen Use in Adults in Healthcare and Emergency Settings. *Thorax* **2017**, *72* (Suppl 1), ii1–ii90. https://doi.org/10.1136/thoraxjnl-2016-209729.

(238) National Institute for Health and Care Excellence. Venous thromboembolism in over 16s: reducing the risk of hospital-acquired deep vein thrombosis or pulmonary embolism. https://www.nice.org.uk/guidance/ng89 (accessed 2022 -01 -31).

(239) Lodha, R.; Kabra, S. K.; Pandey, R. M. Antibiotics for Community-acquired Pneumonia in Children. *Cochrane Database Syst. Rev.* **2013**, No. 6. https://doi.org/10.1002/14651858.CD004874.pub4.

(240) BTS guidelines for the management of community acquired pneumonia in adults: update 2009 | Thorax https://thorax.bmj.com/content/64/Suppl_3/iii1 (accessed 2021 -03 -22).

(241) National Institute for Health and Care Excellence. Pneumonia in adults; Quality standards (QS110) https://www.nice.org.uk/guidance/qs110 (accessed 2021 -08 -20).

(242) Luo, J.; Duke, T.; Chisti, M. J.; Kepreotes, E.; Kalinowski, V.; Li, J. Efficacy of High-Flow Nasal Cannula vs Standard Oxygen Therapy or Nasal Continuous Positive Airway Pressure in Children with Respiratory Distress: A Meta-Analysis. *J. Pediatr.* **2019**, *215*, 199-208.e8. https://doi.org/10.1016/j.jpeds.2019.07.059.

(243) Chu, D. K.; Kim, L. H.-Y.; Young, P. J.; Zamiri, N.; Almenawer, S. A.; Jaeschke, R.; Szczeklik, W.; Schünemann, H. J.; Neary, J. D.; Alhazzani, W. Mortality and Morbidity in Acutely Ill Adults Treated with

Liberal versus Conservative Oxygen Therapy (IOTA): A Systematic Review and Meta-Analysis. *Lancet Lond. Engl.* **2018**, *391* (10131), 1693–1705. https://doi.org/10.1016/S0140-6736(18)30479-3.

(244) Roca, O.; Riera, J.; Torres, F.; Masclans, J. R. High-Flow Oxygen Therapy in Acute Respiratory Failure. *Respir. Care* **2010**, *55* (4), 408–413.

(245) D'Cruz, R. F.; Hart, N.; Kaltsakas, G. High-Flow Therapy: Physiological Effects and Clinical Applications. *Breathe* **2020**, *16* (4). https://doi.org/10.1183/20734735.0224-2020.

(246) Frat, J.-P.; Thille, A. W.; Mercat, A.; Girault, C.; Ragot, S.; Perbet, S.; Prat, G.; Boulain, T.; Morawiec, E.; Cottereau, A.; Devaquet, J.; Nseir, S.; Razazi, K.; Mira, J.-P.; Argaud, L.; Chakarian, J.-C.; Ricard, J.-D.; Wittebole, X.; Chevalier, S.; Herbland, A.; Fartoukh, M.; Constantin, J.-M.; Tonnelier, J.-M.; Pierrot, M.; Mathonnet, A.; Béduneau, G.; Delétage-Métreau, C.; Richard, J.-C. M.; Brochard, L.; Robert, R. High-Flow Oxygen through Nasal Cannula in Acute Hypoxemic Respiratory Failure. *N. Engl. J. Med.* **2015**, *372* (23), 2185–2196. https://doi.org/10.1056/NEJMoa1503326.

(247) Stiller, K. Physiotherapy in Intensive Care: Towards an Evidence-Based Practice. *Chest* **2000**, *118* (6), 1801–1813.

(248) Bott, J.; Blumenthal, S.; Buxton, M.; Ellum, S.; Falconer, C.; Garrod, R.; Harvey, A.; Hughes, T.; Lincoln, M.; Mikelsons, C.; Potter, C.; Pryor, J.; Rimington, L.; Sinfield, F.; Thompson, C.; Vaughn, P.; White, J. Guidelines for the Physiotherapy Management of the Adult, Medical, Spontaneously Breathing Patient. *Thorax* **2009**, *64 Suppl 1*, i1-51.

(249) General Medical Council. Learning disabilities https://www.gmc-uk.org/ethical-guidance/ethical-hub/learning-disabilities (accessed 2021 -08 -20).

(250) Emerson, E.; Baines, S.; Allerton, L.; Welch, V. Health Inequalities & People with Learning Disabilities in the UK: 2012 https://www.basw.co.uk/system/files/resources/basw_14846-4_0.pdf.

(251) Addington-Hall, J.; Kalra, L. Who Should Measure Quality of Life? *BMJ* **2001**, *322* (7299), 1417–1420.

(252) Yates, K.; Festa, M.; Gillis, J.; Waters, K.; North, K. Outcome of Children with Neuromuscular Disease Admitted to Paediatric Intensive Care. *Arch. Dis. Child.* **2004**, *89* (2), 170–175. https://doi.org/10.1136/adc.2002.019562.

(253) Montgomery, J.; Stokes-Lampard, H.; Griffiths, M.; Gardiner, D.; Harvey, D.; Suntharalingam, G. Assessing Whether COVID-19 Patients Will Benefit from Critical Care, and an Objective Approach to Capacity Challenges during a Pandemic: An Intensive Care Society Clinical Guideline. *J. Intensive Care Soc.* **2021**, *22* (3), 204–210. https://doi.org/10.1177/1751143720948537.

(254) Health Education England. The Oliver McGowan Mandatory Training in Learning Disability and Autism. https://www.hee.nhs.uk/our-work/learning-disability/oliver-mcgowan-mandatory-training-learning-disability-autism (accessed 2022 -02 -11).

(255) Forbat, L.; Barclay, S. Reducing Healthcare Conflict: Outcomes from Using the Conflict Management Framework. *Arch. Dis. Child.* **2019**, *104* (4), 328–332. https://doi.org/10.1136/archdischild-2018-315647.

(256) Wallis, C. When Paediatricians and Families Can't Agree. *Arch. Dis. Child.* **2018**, *103* (5), 413–414. https://doi.org/10.1136/archdischild-2017-314414.

(257) General Medical Council. Treatment and Care towards the End of Life Content. GMC 2010.

(258) National Institute for Health and Care Excellence. Improving supportive and palliative care for adults with cancer. CSG4 https://www.nice.org.uk/guidance/csg4 (accessed 2021 -08 -18).

(259) Read, S.; Morris, H. Living and Dying with Dignity: The Best Practice Guide to End-of-Life Care for People with a Learning Disability. Mencap November 1, 2008.

(260) Tuffrey-Wijne, I.; Wicki, M.; Heslop, P.; McCarron, M.; Todd, S.; Oliver, D.; de Veer, A.; Ahlström, G.; Schäper, S.; Hynes, G.; O'Farrell, J.; Adler, J.; Riese, F.; Curfs, L. Developing Research Priorities for Palliative Care of People with Intellectual Disabilities in Europe: A Consultation Process Using Nominal Group Technique. *BMC Palliat. Care* **2016**, *15*, 36. https://doi.org/10.1186/s12904-016-0108-5.

(261) Graham, Y. N.; Hayes, C.; Stephenson, L.; Wanless, A.; Baker, L.; Baker. The Role of Learning Disability Nurses in Providing End-of-Life Care. *Nursing Times*. 2020, pp 18–20.

(262) Romo, R. D.; Lynn, J. The Utility and Value of the "Surprise Question" for Patients with Serious Illness. *CMAJ Can. Med. Assoc. J.* **2017**, *189* (33), E1072–E1073. https://doi.org/10.1503/cmaj.733231.

(263) Toussaint, M.; Boitano, L. J.; Gathot, V.; Steens, M.; Soudon, P. Limits of Effective Cough-Augmentation Techniques in Patients with Neuromuscular Disease. *Respir. Care* **2009**, *54* (3), 359–366.

(264) Mellies, U.; Goebel, C. Optimum Insufflation Capacity and Peak Cough Flow in Neuromuscular Disorders. *Ann. Am. Thorac. Soc.* **2014**, *11* (10), 1560–1568. https://doi.org/10.1513/AnnalsATS.201406-264OC.

(265) Dohna-Schwake, C.; Ragette, R.; Teschler, H.; Voit, T.; Mellies, U. IPPB-Assisted Coughing in Neuromuscular Disorders. *Pediatr. Pulmonol.* **2006**, *41* (6), 551–557.

(266) Jenkins, H. M.; Stocki, A.; Kriellaars, D.; Pasterkamp, H. Breath Stacking in Children with Neuromuscular Disorders. *Pediatr. Pulmonol.* **2014**, *49* (6), 544–553. https://doi.org/10.1002/ppul.22865.

(267) Miske, L. J.; Hickey, E. M.; Kolb, S. M.; Weiner, D. J.; Panitch, H. B. Use of the Mechanical In-Exsufflator in Pediatric Patients With Neuromuscular Disease and Impaired Cough. *Chest* **2004**, *125* (4), 1406–1412.

(268) Veldhoen, E. S.; Verweij-van den Oudenrijn, L. P.; Ros, L. A.; Hulzebos, E. H.; Papazova, D. A.; van der Ent, C. K.; van der Pol, L. W.; Nijman, J.; Wösten-van Asperen, R. M. Effect of Mechanical Insufflation-

Exsufflation in Children with Neuromuscular Weakness. *Pediatr Pulmonol* **2020**, *55* (2), 510–513. https://doi.org/10.1002/ppul.24614.

(269) Chatwin, M.; Ross, E.; Hart, N.; Nickol, A. H.; Polkey, M. I.; Simonds, A. K. Cough Augmentation with Mechanical Insufflation/Exsufflation in Patients with Neuromuscular Weakness. *Eur Respir J* **2003**, *21* (3), 502–508.

(270) 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* (2), 187–193. https://doi.org/10.4187/respcare.05663.

(271) McCarren, B.; Alison, J. A.; Herbert, R. D. Vibration and Its Effect on the Respiratory System. *Aust. J. Physiother.* **2006**, *52* (1), 39–43. https://doi.org/10.1016/s0004-9514(06)70060-5.

(272) Main, E.; Denehy, L. *Cardiorespiratory Physiotherapy: Adults and Paediatrics 5th Edition*; Elsevier, 2016.

(273) Schöni, M. H. Autogenic Drainage: A Modern Approach to Physiotherapy in Cystic Fibrosis. *J. R. Soc. Med.* **1989**, *82* (Suppl 16), 32–37.

(274) King, M.; Phillips, D. M.; Gross, D.; Vartian, V.; Chang, H. K.; Zidulka, A. Enhanced Tracheal Mucus Clearance with High Frequency Chest Wall Compression. *Am. Rev. Respir. Dis.* **1983**, *128* (3), 511–515. https://doi.org/10.1164/arrd.1983.128.3.511.

(275) Yuan, N.; Kane, P.; Shelton, K.; Matel, J.; Becker, B. C.; Moss, R. B. 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* (7), 815–821. https://doi.org/10.1177/0883073809350223.

(276) Lechtzin, N.; Wolfe, L. F.; Frick, K. D. The Impact of High-Frequency Chest Wall Oscillation on Healthcare Use in Patients with Neuromuscular Diseases. *Ann Am Thorac Soc* **2016**, *13* (6), 904–909. https://doi.org/10.1513/AnnalsATS.201509-597OC.

(277) Bidiwala, A.; Volpe, L.; Halaby, C.; Fazzari, M.; Valsamis, C.; Pirzada, M. A Comparison of High Frequency Chest Wall Oscillation and Intrapulmonary Percussive Ventilation for Airway Clearance in Pediatric Patients with Tracheostomy. *Postgrad Med* **2017**, *129* (2), 276–282. https://doi.org/10.1080/00325481.2017.1264854.

(278) Douglas, N. J.; Drummond, G. B.; Sudlow, M. F. Breathing at Low Lung Volumes and Chest Strapping: A Comparison of Lung Mechanics. *J. Appl. Physiol.* **1981**, *50* (3), 650–657. https://doi.org/10.1152/jappl.1981.50.3.650.

(279) Pulletz, S.; Elke, G.; Zick, G.; Schädler, D.; Reifferscheid, F.; Weiler, N.; Frerichs, I. Effects of Restricted Thoracic Movement on the Regional Distribution of Ventilation. *Acta Anaesthesiol. Scand.* **2010**, *54* (6), 751–760. https://doi.org/10.1111/j.1399-6576.2010.02233.x.

(280) Eberlein, M.; Schmidt, G. A.; Brower, R. G. Chest Wall Strapping. An Old Physiology Experiment with New Relevance to Small Airways Diseases. *Ann. Am. Thorac. Soc.* **2014**, *11* (8), 1258–1266. https://doi.org/10.1513/AnnalsATS.201312-465OI.

(281) Mendonca, C. T.; Schaeffer, M. R.; Riley, P.; Jensen, D. Physiological Mechanisms of Dyspnea during Exercise with External Thoracic Restriction: Role of Increased Neural Respiratory Drive. *J. Appl. Physiol. Bethesda Md 1985* **2014**, *116* (5), 570–581. https://doi.org/10.1152/japplphysiol.00950.2013.

(282) Garuti, G.; Verucchi, E.; Fanelli, I.; Giovannini, M.; Winck, J. C.; 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. https://doi.org/10.1186/s13052-016-0216-0.

(283) Holland, A. E.; Denehy, L.; Ntoumenopoulos, G.; Naughton, M. T.; Wilson, J. W. Non-Invasive Ventilation Assists Chest Physiotherapy in Adults with Acute Exacerbations of Cystic Fibrosis. *Thorax* **2003**, *58* (10), 880–884. https://doi.org/10.1136/thorax.58.10.880.

(284) Chatwin, M.; Toussaint, M.; Gonçalves, M. R.; Sheers, N.; Mellies, U.; Gonzales-Bermejo, J.; Sancho, J.; Fauroux, B.; Andersen, T.; Hov, B.; Nygren-Bonnier, M.; Lacombe, M.; Pernet, K.; Kampelmacher, M.; Devaux, C.; Kinnett, K.; Sheehan, D.; Rao, F.; Villanova, M.; Berlowitz, D.; Morrow, B. M. Airway Clearance Techniques in Neuromuscular Disorders: A State of the Art Review. *Respir. Med.* **2018**, *136*, 98–110. https://doi.org/10.1016/j.rmed.2018.01.012.

(285) Sue, K.; Mazzotta, P.; Grier, E. Palliative Care for Patients with Communication and Cognitive Difficulties. *Can. Fam. Physician* **2019**, *65* (Suppl 1), S19–S24.

(286) General Medical Council. Decision Making and Consent. GMC 2020.

(287) NHS England. Delivering High Quality End of Life Care for People Who Have a Learning Disability. NHS August 16, 2017.

(288) Tuffrey-Wijne, I.; Davidson, J. Excellence in Palliative and End-of-Life Care Provision for People with Intellectual Disability. *Int. J. Palliat. Nurs.* **2018**, *24* (12), 598–610. https://doi.org/10.12968/ijpn.2018.24.12.598.

(289) National Institute for Health and Care Excellence. End of Life Care for Infants, Children and Young People with Life-Limiting Conditions: Planning and Management. NICE 2016.

(290) Givens, J. L.; Jones, R. N.; Shaffer, M. L.; Kiely, D. K.; Mitchell, S. L. Survival and Comfort After Treatment of Pneumonia in Advanced Dementia. *Arch. Intern. Med.* **2010**, *170* (13), 1102–1107. https://doi.org/10.1001/archinternmed.2010.181.

(291) Ahronheim, J. C.; Morrison, R. S.; Baskin, S. A.; Morris, J.; Meier, D. E. Treatment of the Dying in the Acute Care Hospital. Advanced Dementia and Metastatic Cancer. *Arch. Intern. Med.* **1996**, *156* (18), 2094–2100.
(292) Rosenberg, J. H.; Albrecht, J. S.; Fromme, E. K.; Noble, B. N.; McGregor, J. C.; Comer, A. C.; Furuno, J. P. Antimicrobial Use for Symptom Management in Patients Receiving Hospice and Palliative Care: A Systematic Review. *J. Palliat. Med.* **2013**, *16* (12), 1568–1574. https://doi.org/10.1089/jpm.2013.0276.
(293) British Medical Association; Royal College of Physicians; General Medical Council. Clinically-Assisted Nutrition and Hydration (CANH) and Adults Who Lack the Capacity to Consent. Guidance for Decision-Making in England and Wales. British Medical Association 2018.
(294) Regnard, C.; Reynolds, J.; Watson, B.; Matthews, D.; Gibson, L.; Clarke, C. Understanding Distress in People with Severe Communication Difficulties: Developing and Assessing the Disability Distress Assessment Tool (DisDAT). *J. Intellect. Disabil. Res.* **2007**, *51* (4), 277–292. https://doi.org/10.1111/j.1365-2788.2006.00875.x.
(295) Baillie, J.; Anagnostou, D.; Sivell, S.; Van Godwin, J.; Byrne, A.; Nelson, A. Symptom Management, Nutrition and Hydration at End-of-Life: A Qualitative Exploration of Patients', Carers' and Health Professionals' Experiences and Further Research Questions. *BMC Palliat. Care* **2018**, *17* (1), 60. https://doi.org/10.1186/s12904-018-0314-4.
(296) Hunt, K.; Bernal, J.; Worth, R.; Shearn, J.; Jarvis, P.; Jones, E.; Lowe, K.; Madden, P.; Barr, O.; Forrester-Jones, R.; Kroll, T.; McCarron, M.; Read, S.; Todd, S. End-of-Life Care in Intellectual Disability: A Retrospective Cross-Sectional Study. *BMJ Support. Palliat. Care* **2020**, *10* (4), 469–477. https://doi.org/10.1136/bmjspcare-2019-001985.
(297) Selman, L. E.; Chao, D.; Sowden, R.; Marshall, S.; Chamberlain, C.; Koffman, J. Bereavement Support on the Frontline of COVID-19: Recommendations for Hospital Clinicians. *J. Pain Symptom Manage.* **2020**, *60* (2), e81–e86. https://doi.org/10.1016/j.jpainsymman.2020.04.024.

10. Acknowledgements

Professor A John Simpson is a National Institute for Health Research (NIHR) Senior Investigator. The views expressed in this article are those of the author(s) and not necessarily those of the NIHR, or the Department of Health and Social Care.
We are grateful to Drs Akshay Batra and Emma Jones, University Hospitals Southampton, for their review of elements of the manuscript relating to gastro-oesophageal reflux.
The authors would like to thank Katy Kerr, Health Education England, for her help and guidance relating to the oral health sections of the statement.

11. Disclosures

Michelle Chatwin discloses that her clinical practice is at the Royal Brompton Hospital and National Hospital for Neurology and Neurosurgery, University College Hospital. She also works part time developing the education and research programme at Breas Medical. Breas Medical have had no influence or part in the writing of this manuscript.

2239

Appendix 1 Airway Clearance Techniques

Clearance Technique Name	Description	Advantages	Disadvantages	Notes and Evidence Base
Manual Assisted Cough (MAC)	A MAC can be delivered to the abdomen and / or the thorax. Compression of the abdomen causes a sudden increase in abdominal pressure; this causes the abdominal contents to push the diaphragm upwards, increasing expiratory airflow. Similarly, sudden thoracic compression causes air to be rapidly expelled, with acceleration of airflow towards the mouth. The technique involves the patient taking a spontaneous, or receiving an assisted, inspiration and at the start of the cough expiratory compression is applied.	The technique is easy to perform and can be utilised in multiple settings and in conjunction with assisted inspiration and Mechanical Insufflation-Exsufflation	Requires co-ordination between the carer delivering the technique and the individual receiving it. May not be effective in individuals with a very weak cough. May be less effective in patients with severe scoliosis	Simple effective technique. Been shown to increase CPF in children and adults with NMD compared with their unassisted CPF[263]
Assisted inspiration with either non-invasive ventilation (NIV), intermittent positive pressure breathing (IPPB), resuscitation bag or lung volume recruitment circuit (LVR)	Single breath assisted inspiration provides a single, sustained inspiratory flow that inflates the respiratory system to the maximal desired volume. Once this volume has been attained, the patient breathes out or coughs. The use of a resuscitation bag or LVR circuit enables repeated inspirations if the patient can breath hold (air stacking (AS)). Both these techniques increase lung volumes and expiratory flows, with repeated assisted inspiration having the potential to increase this the most.	The technique is easy to perform and can be utilised in multiple settings. Can be used with a MAC to increase efficacy further.	Some patients have difficulty accepting extrinsic breaths. When using multiple assisted inspirations the patient must co-ordinate with the technique and be able to signal when they have received sufficient inspiratory volume. This may not be possible in those with severe Lerning Disability.	Assisted inspiration increases CPF and lung volumes in children[264,265]. Jenkins et al. investigated 23 children's ability to learn AS using a LVR circuit, eight of whom had some degree of Learning disability. Only four participants were unable to effectively AS[266].
Mechanical Insufflation-Exsufflation (MI-E)	MI-E is a device that applies a positive pressure to the airway (insufflation) followed by a rapid switch to negative pressure (exsufflation), aiming to simulate the natural flow changes that occur with a cough.	Increases cough efficacy in the weakest patients (CPF <160L/min). Has the potential to shift a large quantity of secretions.	MI-E can be difficult to perform in very young infants who are unable to accept insufflation.	MI-E has been shown to increase CPF, decrease rates of CAP hospitalisations and reduce length of hospital stay in NMD patients[267–269]. In patients with CP, MI-E did not show any benefit over conventional physiotherapy in length of hospital stay or days on oxygen in a randomised trial of 22 patients admitted with CAP[270].

2240
2241

Table I Proximal Airway Clearance Techniques. Abbreviations: AS= Air Stacking; CAP= Community Acquired Pneumonia; CPF= Cough Peak Flow; IPPB= Intermittent Positive Pressure Breathing; LVR= Lung Volume Recruitment; MAC=Manual Assisted Cough; MI-E= Mechanical Insufflation-Exsufflation; NIV= Non-Invasive Ventilation; NMD= Neuromuscular Disorder

Peripheral Airway Clearance Technique Name	Typical Treatment Time	Description	Advantages	Disadvantages	Evidence Base
Conventional physiotherapy (CPT) (postural drainage and percussion)	5 minutes each lung area	Manual techniques consist of chest percussion and vibrations or shaking. This is performed using a hand, fingers or facemask and is generally well tolerated. Chest vibrations consist of a rapid extra-thoracic force at the beginning of expiration, followed by oscillatory compressions until expiration is complete [271]. This is believed to increase peak expiratory flow.	Can be performed in patients that cannot co-operate with other techniques. Can be combined with other techniques to enhance airway clearance. Suitable for all ages. Easy to teach.	May not be effective when used in isolation. Care needs to be taken when applying shaking and vibrations not to take the patient to lung closing volumes.	Despite the lack of evidence base this can be effective in clearing secretions. An ENMC workshop report and review recommended the use of these techniques in patients with NMD [75].
Positioning / gravity assisted postural drainage	5 minutes each lung area	The technique involves placing the patient in a position which drains mucus from the periphery of the lungs to the central upper airway. The addition of gravity can assist in the drainage of secretions further.	Any patient can be positioned to utilise this technique.	The addition of gravity may not be appropriate for all patients especially those who have severe gastroesophageal reflux disease.	Despite the lack of evidence, clinically this can be effective in clearing secretions.
Breathing exercises- active cycle of breathing technique (ACBT) and autogenic drainage (AD)	5-10 minutes	ACBT uses a cycle of techniques to loosen airway secretions by increasing tidal volumes, increasing expiratory airflow and utilising channels of co-lateral ventilation. The technique consists of breathing control, thoracic expansion exercises, and the forced expiration technique[272]. AD consists of breathing at different lung volumes to enable "unsticking", "collecting" and "evacuating" of secretions. The aim is to achieve high expiratory flows by narrowing the airway through an open glottis whilst forcing expiration after taking a slow deep inspiration with an inspiratory hold.[273]	The techniques do not require any equipment.	These techniques are difficult to learn if the patient has cognitive impairment or is unable to participate. Patients with respiratory muscle weakness also find them difficult to perform if they are unable to independently take a deep breath in.	There is no evidence base for these techniques in learning disability and neurodisability. However, may be effective in patients without weakness and who are able to execute the techniques adequately. .
Positive expiratory pressure (PEP) and oscillatory PEP	5 to 10 breaths 5-10 minutes	PEP involves breathing out against a set resistance through a mouthpiece or mask. It prevents airway collapse, utilises collateral channels of ventilation, decreases hyperinflation and increases lung volumes. Oscillatory PEP has the same principles as PEP but with the addition of oscillations to create internal vibrations within the airway.	The equipment is not expensive and can be used in any environment. For the most effective results there does need to be some co-operation from the patient	This technique can induce further fatigue in patients that are weak. It can also be ineffective in patients with low expiratory flows	There is no evidence base for these techniques in learning disability and neurodisability. However, may be effective in patients without weakness and who are able to execute the techniques adequately.

Oscillatory devices: high frequency chest wall oscillation (HFCWO) and intrapulmonary percussive ventilation (IPV)	10-30 minutes	HFCWO provides compression of the chest wall at frequencies that are similar to the resonant frequency of the lung, between 5-20 Hz [274], via an air pulse generator then delivers intermittent positive airflow into the jacket. As the jacket expands compressing the chest wall, it produces a transient / oscillatory increase in airflow in the airways vibrating and creating a expiratory air flow bias moving the secretions from the peripheral airways toward the mouth. IPV is delivered via an IPPB pneumatic device. IPV delivers air to the lungs at frequencies of 100 to 300 cycles per minute at peak pressures from 10 to 40 cmH2O. IPV superimposes high-frequency bursts of gas on top of the patient's own respiration. This creates a global effect of internal percussion of the lungs, which promotes clearance from the peripheral bronchial tree. The high frequency airflow pulsates to expand the lungs, vibrate and enlarge the airways. This delivers air to the distal lung units, beyond accumulated secretions.	HFCWO can be used in conjunction with ventilator support. IPV can also provide ventilatory support. Neither device requires co-operation from the patient. The vibration and movement from the secretions with this technique can elicit a spontaneous cough in individuals who have desensitised their cough reflex.	IPV is not available at present within the UK. HFCWO devices currently require individual funding requests in UK.	IPV and HFCWO have been shown to decrease hospitalisations and requirement of antibiotics for CAP in neurodisability [275–277].
Chest wall strapping (CWS)	unclear	CWS is the restriction of the chest wall through the application of elastic material around the thorax. Strapping via CWS passively lowers the functional residual capacity (FRC) without using expiratory muscles. This has been demonstrated to be beneficial for lung secretion clearance [278]. The principles and physiological effects of CWS are similar to that of Autogenic Drainage [273]. The single most important physiologic change consists of the significant increase in maximal expiratory flow.	Simple effective treatment that does not require co-operation of the patient. If the patient is fatigued, can be used in conjunction with ventilatory support.	Requires a skilled therapist to teach the technique and assess whether it needs to be used in conjunction with other techniques.	There is no evidence regarding deflation and strapping to mimic breathing at a low lung volume as in the airway clearance technique, autogenic drainage (deflation). However, physiological arguments and clinical experience advocate for using CWS which induces breathing at low lung volumes, increases lung elastic recoil and increases maximal expiratory flows [279–281].
Expiratory flow acceleration (EFA)	10-30 minutes	EFA is a technique that increases expiratory flow and enhances natural secretion movement.	A simple treatment that requires no co-operation from the patient and there are no contraindications in the spontaneously breathing patient.	A device is required to provide this treatment.	EFA has been shown found in an observational study to reduce primary care visits and hospitalisation for CAP in CP patients [282].
Non-invasive ventilation (NIV) and intermittent positive pressure breathing (IPPB)	10-30 minutes	NIV and IPPB provide ventilatory support set to provide a greater tidal volume than they can manage independently. Increasing tidal volume increases the expiratory airflow bias and utilises collateral channels of ventilation to enhance secretion movement.	It is not necessary for patient co-operation, can off load the work of breathing and prevent oxygen desaturation during treatment.	A device is required to provide this treatment.	NIV is a recognised tool for airway clearance in adults and children with cystic fibrosis[283]. It has been recommended for use in patients with NMD [75,284].

Table II Peripheral Airway Clearance Techniques Abbreviations: ACBT= Active Cycle of Breathing Technique; AD= Autogenic Drainage; CAP= Community Acquired Pneumonia; CP= Cerebral Palsy; CPT= Conventional PhysioTherapy; CWS= Chest Wall Strapping; EFA= Expiratory Flow Acceleration; ENMC= European Neuromuscular Centre; FRC= Functional Residual Capacity; HFCWO= High Frequency Chest Wall Oscillation; IPPB= Intermittent Positive Pressure Breathing; IPV= Intrapulmonary Percussive Ventilation; NIV= Non-Invasive Ventilation; NMD= Neuromuscular Disease; PEP= Positive Expiratory Pressure;

Appendix 2 – Palliative Care Considerations

Breaking bad news

Breaking bad news to patients with learning disability requires experience and an understanding of their capacity. The ARCH model [259] was developed as a specific framework for breaking bad news to people with learning disability, particularly focused on helping carers. It suggests considering four parts: Ask, Repeat and clarify, Check level of understanding, Help person to express feelings.

Breaking bad news should not be avoided because someone has a learning disability. Supporting them in understanding what is happening to them can improve quality of life and reduce anxiety at the end of life [285].

Legal aspects in decision making

All people with a learning disability should be involved in making decisions about their care to the extent they are able. The presumption in treating all patients is that they have capacity to make decisions, however, if this is in doubt, their capacity should be formally assessed following country-specific guidance [286]. Mental capacity is both time and decision-specific. If the patient does not have capacity, to make a particular treatment decision, the best course of action may be determined by the medical team in conjunction with the patient's family/friends on a 'best-interests' basis. This will consider the patient's previously expressed values/preferences, and whether anybody else has the legal authority to make decisions on the patient's behalf.

If the patient previously had capacity, they may have chosen to nominate a trusted family member/friend to make healthcare decisions on their behalf. (Lasting Power of Attorney (LPA) for Health and Welfare in England and Wales or Welfare Power of Attorney in Scotland). In the event that the patient loses capacity to make relevant decisions, this grants decision-making power to the elected attorney.

If the patient does not have capacity and has no LPA, or has never had capacity, applications can be made for a deputy (England and Wales) or Guardian (Scotland) to be appointed. However, an appointed deputy may not make decisions to discontinue life-sustaining treatment.

Advance care plans

Advance care plans are a formal care plan that includes details about the patient's condition, decisions made with them and their family/carers (for example about managing symptoms), and their wishes and ambitions. This plan is a core element of their palliative care. A healthcare directive or advance care plan (ACP) may be useful in patients (with mental capacity) to support them in discussing and making decisions about their future health [10,287,288].

It is recognised that patients with learning disability are less likely to have had these discussions, and as such, more likely to have a poorly coordinated death which was less likely to have specialist palliative care input [10]. More comprehensive ACPs may include details about parallel planning and this can be important in recognising the unpredictability of life-limiting conditions, and making multiple plans of care for different eventualities [289] including treatment of future pneumonias. ACPs may also advocate for further active interventions. Advance Decision to Refuse Treatment (ADRT) are legally binding documents which can be tailored by the patient to specify exactly what treatments they would not wish to have in specific situations. These are best completed with legal advice to ensure clarity in documentation.

To support decision-making and weigh up benefits and burdens of treatment, quality of life must be considered. This can only be truly assessed by the individual. It may be that a patient chooses to focus on aspects that are important to their quality of life such that they make choices that may pose further risk to their health (e.g. continuing oral diet despite evolving bulbar dysfunction). This may be acceptable if the patient has capacity and has been fully counselled as to the ongoing risks.

Making a judgement on a patient's quality of life without full assessment risks providing inadequate, impersonal care. Where a patient is unable to communicate their quality of life, proxies are almost as good at detecting changes in quality of life as the individual [251] where health professionals may underestimate it.

For those lacking mental capacity, an ACP can still be useful to reflect the preferences and recommendations from the family/friends and clinical teams that know them.

Treatment of infections in EOL

Palliative care is focused on improving quality of life and weighing up the benefits and burdens of further intervention/treatment.

It is particularly important to review both the benefits and burdens of investigations and treatments when a patient is reaching the end of their life. In the last few days of life for example, it may be that there is no overall benefit in continuing potentially life prolonging but burdensome treatment [257].

Decisions to continue with intravenous (or other burdensome) medications when a patient is at an advanced stage may impede significantly on their quality of life, particularly if it limits their choice of preferred place of care. There are some suggestions that continuation of antibiotics (oral or intravenous) may even prolong the dying process and reduce comfort [290]. Others hypothesise that development of a serious infection, left untreated, leading to sedation may provide for a peaceful death [291]. Conversely, antibiotics in end of life have also been associated with improved symptom management [292].

Ultimately this comes down to communication with the patient (or proxy) to encourage shared and informed decision making. Ideally, these discussions would have already happened as part of advance care planning.

Clinically Assisted Nutrition and Hydration in EOL

Clinically assisted nutrition and hydration (CANH) may need reassessment at end of life. Individualised decisions should be made weighing up benefits and burden, and based on the patient's and family's understanding, concerns and wishes around food and fluid. As a medical intervention, it is legal/ethical to withdraw/withhold CANH at the end of life. There is clear guidance on the management of this at end of life in those lacking capacity [257,293]. In the last few days of life, it is unlikely that CANH will improve symptoms or prolong life but may cause additional discomfort.

Symptom management

Patients with learning disability may not be able to clearly communicate their symptom burden. It is essential that someone who knows the patient is involved in guiding the clinician in assessing symptoms and distressing behaviours. Using a tool like the Disability Distress Assessment Tool (DisDAT) can be beneficial in making this assessment [294]. There are also pain tools that can be useful

in assessing non-verbal patients such as Wong-Baker FACES pain rating scale, Visual Analog Scale and FLACC.

Symptom management should be tailored to the patient, their comorbidities, and symptoms. This can be complex and the benefit of a local palliative care team advising on and anticipating the management of symptoms cannot be underestimated [257,295]. Non-pharmacological methods should always be considered alongside pharmacological methods. Clear, honest and open communication can reduce anxiety and symptom burden in end-of life. As part of symptom management, it is important to reassess the need for interventions (e.g. blood tests), and to consider rationalising the medication burden.

Preferred place of care

Preferred place of care and place of death may have been discussed as part of an ACP. It is important to recognise that where possible patients should be cared for where they are likely to be most comfortable for end-of-life care. Options may include home, hospital, hospice or another residential setting. Although home is often considered to be the most desired place, it may not be right in all situations so other places should be explored. It must be noted that to support patients with learning disability who die in residential care settings may require increased input and support for carers who may not be experienced in end-of-life care [288]. Carers in residential settings report that they often feel unheard in decision-making about their clients [288].

Bereavement support

Bereavement support is a continuing aspect of palliative care. Hospital bereavement services, GPs and specialist palliative care teams will be able to provide guidance on where the family/friends of the deceased can access this. Consideration must also be given to how to support carers and other residents in residential settings with bereavement, particularly when the person dies in another setting [296]. Bereavement distress can be mitigated with clear communication with family/carers – updating them on the patient's condition particularly though honest discussions when death is anticipated [297]. Families also value staff making contact with them after death e.g. through a condolence letter/card or offering a follow up appointment to answer any questions they may have.