

APPENDIX 1. CAUSES OF ABNORMAL SWALLOWING

Abnormalities of the timing and amplitude of laryngeal movement reduce the duration and extent of upper oesophageal sphincter opening, which leads to pooling of oral secretions. New or pre-existing neurological disorders may drive such abnormalities, as may iatrogenic interventions such as botulinum toxin to the sternomastoid muscles.

A common upper motor neurone (UMN) cause of swallowing failure is middle cerebral artery (MCA) stroke. Fine supranuclear control of intra-oral bolus preparation, control, and propulsion is disrupted without denervation of the larynx, palate, face, pharynx, and tongue. Recovery can be relatively prompt (within a couple of weeks) partly because reflex swallowing may be retained.

Muscular dystrophies, particularly oculopharyngeal muscular dystrophy, are characterised by profound and selective weakness without any central or peripheral cranial nerve palsy. In contrast to MCA stroke, preservation of UMN control means that, strength permitting, the bolus can be prepared with reasonable precision and initiation of bolus propulsion is under voluntary control. In children with neuromuscular diseases abnormal swallowing (47%) and dysarthria (32%) were common (i). Duchenne's muscular dystrophy (DMD) is the most common muscular dystrophy and DMD Pathfinders has developed useful guidance for managing nutrition, including daily living tips and recipes to help people with DMD with various degrees of swallowing difficulties (ii). Consensus guidelines on adults with DMD recommend that consideration of swallowing and nutritional status be included in regular assessment of patients (iii).

Motor neurone disease (MND) has a variable upper and lower motor neurone pattern, accompanied by fronto-temporal dementia in some patients, which can lead to an impulsive and reckless voluntary swallow. Patients with UMN-dominant MND may retain an intact laryngeal reflex, promoting powerful cough responses, even with modest respiratory function.

The oesophageal phase of swallowing may be disrupted by oesophageal dysmotility disorders such as achalasia, which is characterised by failure of relaxation of the lower oesophageal sphincter and ineffective peristalsis (iv). Mechanical obstruction also explains the association between AP and malignant or benign oesophageal strictures. The risk of AP increases with stricture severity. Eosinophilic oesophagitis is the second most common benign cause of oesophageal swallowing difficulty and is associated with AP (v,vi).

In patients with oesophageal cancer, tracheo-oesophageal or broncho-oesophageal fistulae can produce AP. Upper GI endoscopy, and in particular emergency endoscopy with or without prophylactic intubation, can also lead to AP (vii,viii,ix). Furthermore, the profound cachexia and sarcopenia that may accompany chronic abdominal conditions such as cirrhosis can drive abnormal swallowing and AP (x).

APPENDIX 2. SPECIFIC ADDITIONAL CONSIDERATIONS IN PALLIATIVE CARE

Decisions around eating and drinking with acknowledged risks

At the end of life, some patients may choose to accept the risk of continuing to eat and drink by mouth, or this may be discussed and decided to be in their best interest following legally-guided mental capacity processes. SLT and dietetic input is essential to support these decisions (xi). For some patients the desire to continue to eat orally is driven by the sensory and social enjoyment of eating. By understanding the individual's priorities and preferences, it may be possible to replicate some aspects of mealtimes that are enjoyable in as safe a way as possible.

When a patient chooses to continue to eat or drink, support and advice should be offered to enable them to do this as safely as possible. The Royal College of Physicians has issued helpful guidance around this (xii).

Symptom management

The Leadership Alliance for Care of Dying People highlighted that, at the end of life, we have “one chance to get it right” (xiii). Guidance for management of respiratory symptoms at the end of life has been developed for adults and children (xiv,xv).

Regular assessment should be made of common physical symptoms including cough, breathlessness, excess secretions, delirium, fever, fatigue, pain and anxiety. Symptom management should incorporate non-pharmacological and pharmacological interventions as advocated by local and national guidelines. Response to treatment should be reviewed regularly as titration may be needed. There is limited evidence relating specifically to symptom management in AP, however generic management of some of the most troublesome symptoms related to AP at the end of life are considered in Table i.

Bereavement support

Bereavement can have a significant impact on the psychological and physical health of those affected. It is important to offer information about bereavement support to the people who are affected by the patient's death. This may include professional carers and co-residents if the patient lived in a residential or care home, as well as friends and family (xvi).

Cough and secretions

Opioids have been shown to suppress cough and reduce the related pain and distress. Where excess saliva production is the predominant problem, antimuscarinic drugs are frequently used to reduce the volume, which may relieve cough in some. Although evidence from randomised clinical trials is limited (xvii,xviii) a trial of hyoscine (in either butylbromide or hydrobromide form), glycopyrronium or sublingual atropine ophthalmic solution may be appropriate (xix). It is important to note that antimuscarinic drugs are less effective on purulent bronchial secretions, and in some situations may worsen symptoms by increasing the viscosity of secretions, making them more difficult or distressing to clear. In this situation, nebulised saline may be more effective.

Physiotherapy may also be helpful, as described in section 6, if this is tolerable to the patient (xx).

“Death rattle”

At the end of life, an unconscious patient may develop audible wet sounding breathing due to the reduced ability to sense or clear secretions. The patient is unlikely to be distressed by this, but it can be disturbing to those at the bedside.

If the patient remains comfortable with no signs of distress, no treatment beyond good mouthcare is needed. Positional changes or suctioning may help, but there is little evidence for most pharmacological interventions (xxi,xxii). It is important to give reassurance to the patient’s family and friends, preparing them for normal changes in the sound and rhythm of breathing as the patient reaches the end of life. If the patient is distressed and has profuse secretions, an antimuscarinic drug may reduce excess saliva production (xxiii), however treatment should be stopped if it is ineffective.

Breathlessness

Non-pharmacological approaches to dyspnoea that are supported by evidence aim to:

- Improve the efficiency of the breathing cycle (use of a hand-held fan, optimal positioning and breathing techniques).
- Address cognitive factors that exacerbate breathlessness (relaxation, psychology support).
- Optimise the patient’s ability to function (walking aids, rehabilitation) (xxiv).

Low dose morphine has been shown to reduce the intensity of breathless in advanced disease without leading to clinically significant respiratory adverse effects (xxv,xxvi). A low dose of sustained release opioid is safe and effective in relieving breathlessness (xxvii).

While benzodiazepines are often used to relieve breathlessness there is little evidence to support this, and their effectiveness is related to reducing anxiety. They may have a role if breathlessness persists despite a combination of opioids and non-pharmacological approaches (xxviii).

There is limited evidence that supplementary oxygen improves breathlessness in patients requiring palliative care. It has been recommended that oxygen be considered in people with

breathlessness who have symptomatic hypoxaemia, but only alongside the other measures described above (xxix). Symptoms correlate poorly with blood oxygen levels so assessment of efficacy should be based on relief of symptoms rather than increasing oxygen levels. There is emerging interest in the potential role of non-invasive ventilation and high flow nasal oxygen in relieving breathlessness in some palliative patients (xxx-xxxiii). Decisions around commencing these interventions should take into account the patient's priorities for place of care as well as any possible burdens to the patient, and should always include plans to review benefit, and the circumstances under which the intervention should be discontinued.

Agitation at the end of life

Where severe agitation is causing distress at the end-of-life despite addressing all reversible factors, benzodiazepines (e.g., midazolam or lorazepam) or antipsychotics (e.g., haloperidol or levomepromazine) may be needed (xxxiv,xxxv). These drugs should be introduced at low doses and titrated against response, with the aim of relieving distress rather than inducing sedation. However, the latter may be unavoidable in some circumstances.

Support for the patient's family and friends is essential, since agitation and delirium can be distressing.

Table i. Symptom management at the end of life.

APPENDIX REFERENCES

- i. Kooi-van Es M, Erasmus CE, de Swart BJM, Voet NBM, van der Wees PJ, de Groot IJM, van den Engel-Hoek L, studygroup Dutch pediatric rehabilitation centers. Dysphagia and dysarthria in children with neuromuscular diseases, a prevalence study. *J Neuromuscul Dis* 2020;7:287-95.
- ii. <https://dmdpathfinders.org.uk/resources/advice-guides/>
- iii. Quinlivan R, Messer B, Murphy P, Astin R, Mukherjee R, Khan J, Emmanuel A, Wong SC, Kulshresha R, Willis T, Pattni J, Willis D, Morgan A, Savvatis K, Keen R, Bourke J, Marini Bettolo C, Hewamadduma C, Adult North Star Network. Adult North Star Network (ANSN): consensus guideline for the standard of care of adults with Duchenne muscular dystrophy. *J Neuromuscul Dis* 2021;8:899-926.
- iv. Harvey PR, Coupland B, Mytton J, Evison F, Patel P, Trudgill NJ. Outcomes of pneumatic dilatation and Heller's myotomy for achalasia in England between 2005 and 2016. *Gut* 2019;68:1146-51.
- v. Moawad FJ. Eosinophilic esophagitis: incidence and prevalence. *Gastrointest Endosc Clin N Am* 2018;28:15-25.
- vi. Attwood S. Eosinophilic oesophagitis – a common disease, newly recognised. *Clin Med (Lond)* 2013;13 (Suppl 6):s32-s35.
- vii. Liebler JM, Benner K, Putnam T, Vollmer WM. Respiratory complications in critically ill medical patients with acute upper gastrointestinal bleeding. *Crit. Care Med.* 1991; 19: 1152–7.

- viii. Chaudhuri D, Bishay K, Tandon P, Trivedi V, James PD, Kelly EM, Thavorn K, Kyeremanteng K. Prophylactic endotracheal intubation in critically ill patients with upper gastrointestinal bleed: a systematic review and meta-analysis. *JGH Open*. 2019;24:4:22-8.
- ix. Kawanishi K, Kato J, Toda N, Yamagami M, Yamada T, Kojima K, Ohki T, Seki M, Tagawa K. Risk factors for aspiration pneumonia after endoscopic hemostasis. *Dig Dis Sci*. 2016;61:835-40.
- x. Haraguchi M, Miyaaki H, Nakamura Y, Narita S, Matsumoto K, Fukushima M, Sasaki R, Miuma S, Takahata H, Yamaguchi N, Nakao K. Assessment of the association between dysphagia and sarcopenia among elderly patients with cirrhosis: usefulness of the finger-ring test. *Arch Gerontol Geriatr* 2021;95:104430.
- xi. Soar N, Birns J, Sommerville P, Lang A, Archer S. Approaches to eating and drinking with acknowledged risk: a systematic review. *Dysphagia* 2021;36:54-66.
- xii. Royal College of Physicians. 2021. Supporting people who have eating and drinking difficulties. A guide to practical care and clinical assistance, particularly towards the end of life. Report of a working party. London: RCP.
- xiii. Leadership Alliance for Care of Dying People. One chance to get it right. London: UK Government, 2014.
- xiv. Craig F, Henderson EM, Bluebond-Langner M. Management of respiratory symptoms in paediatric palliative care. *Curr Opin Support Palliat Care* 2015;9:217-26.
- xv. Booth S, Johnson MJ. Improving the quality of life of people with advanced respiratory disease and severe breathlessness. *Breathe (Sheff)* 2019;15:198-215.
- xvi. 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;10:469-77.
- xvii. Squires N, Wills A, Rowson J. The management of drooling in adults with neurological conditions. *Curr Opin Otolaryngol Head Neck Surg* 2012;20:171-6.
- xviii. Walshe M, Smith M, Pennington L. Interventions for drooling in children with cerebral palsy. *Cochrane Database Syst Rev* 2012 Nov 14;11:CD008624.
- xix. McGeachan AJ, McDermott CJ. Management of oral secretions in neurological disease. *Pract Neurol* 2017;17:96-103.
- xx. Arcuri JF, Abarshi E, Preston NJ, Brine J, Pires Di Lorenzo VA. Benefits of interventions for respiratory secretion management in adult palliative care patients - a systematic review. *BMC Palliat Care* 2016 Aug 9;15:74.
- xxi. Wee B, Browning J, Adams A, Benson D, Howard P, Klepping G, Molassiotis A, Taylor D. Management of chronic cough in patients receiving palliative care: review of evidence and recommendations by a task group of the Association for Palliative Medicine of Great Britain and Ireland. *Palliat Med* 2012;26:780-7.
- xxii. Kolb H, Snowden A, Stevens E. Systematic review and narrative summary: treatments for and risk factors associated with respiratory tract secretions (death rattle) in the dying adult. *J Adv Nurs* 2018;74:1446-62.
- xxiii. Mercadante S. Death rattle: critical review and research agenda. *Support Care Cancer* 2014;22:571-5.
- xxiv. Spathis A, Booth S, Moffat C, Hurst R, Ryan R, Chin C, Burkin J. The Breathing, Thinking, Functioning clinical model: a proposal to facilitate evidence-based breathlessness management in chronic respiratory disease. *NPJ Prim Care Respir Med* 2017;27:27.
- xxv. Barnes H, McDonald J, Smallwood N, Manser R. Opioids for the palliation of refractory breathlessness in adults with advanced disease and terminal illness. *Cochrane Database Syst Rev* 2016 March 31;3:CD011008.
- xxvi. Verberkt CA, van den Beuken-van Everdingen MHJ, Schols JMGA, Datla S, Dirksen CD, Johnson MJ, van Kuijk SMJ, Wouters EFM, Janssen DJA. Respiratory adverse effects of opioids for breathlessness: a systematic review and meta-analysis. *Eur Respir J* 2017 Nov 22;50:1701153.

- xvii. Currow DC, McDonald C, Oaten S, Kenny B, Allcroft P, Frith P, Briffa M, Johnson MJ, Abernethy AP. Once-daily opioids for chronic dyspnea: a dose increment and pharmacovigilance study. *J Pain Symptom Manage* 2011;42:388-99.
- xxviii. Simon ST, Higgins IJ, Booth S, Harding R, Weingärtner V, Bausewein C. Benzodiazepines for the relief of breathlessness in advanced malignant and non-malignant diseases in adults. *Cochrane Database Syst Rev* 2016 Oct 20;10:CD007354.
- xxix. Hardinge M, Annandale J, Bourne S, Cooper B, Evans A, Freeman D, Green A, Hippolyte S, Knowles V, MacNee W, McDonnell L, Pye K, Suntharalingam J, Vora V, Wilkinson T. British Thoracic Society guidelines for home oxygen use in adults. *Thorax* 2015;70 (Suppl 1):i1-i43.
- xxx. Wilson ME, Majzoub AM, Dobler CC, Curtis JR, Nayfeh T, Thorsteinsdottir B, Barwise AK, Tilburt JC, Gajic O, Montori VM, Murad MH. Noninvasive ventilation in patients with do-not-intubate and comfort-measures-only orders: a systematic review and meta-analysis. *Crit Care Med* 2018;46:1209-16.
- xxxi. Ruangsomboon O, Dorongthom T, Chakorn T, Monsomboon A, Praphruetkit N, Limsuwat C, Surabengawong U, Riyapan S, Nakornchai T, Chaisirin W. High-flow nasal cannula versus conventional oxygen therapy in relieving dyspnea in emergency palliative patients with do-not-intubate status: a randomized crossover study. *Ann Emerg Med* 2020;75:615-26.
- xxxii. Ricard JD, Roca O, Lemiale V, Corley A, Braunlich J, Jones P, Kang BJ, Lellouche F, Nava S, Rittayamai N, Spoletini G, Jaber S, Hernandez G. Use of nasal high flow oxygen during acute respiratory failure. *Intensive Care Med* 2020;46:2238-47.
- xxxiii. Mercadente S, Adile C, Ferrera P, Giuliana F, Grassi Y. High-flow nasal oxygen therapy. *BMJ Support Palliat Care* 2022 Mar 24;bmjspcare-2022-003606.
- xxxiv. Bush SH, Leonard MM, Agar M, Spiller JA, Hosie A, Wright DK, Meagher DJ, Currow DC, Bruera E, Lawlor PG. End-of-life delirium: issues regarding recognition, optimal management, and the role of sedation in the dying phase. *J Pain Symptom Manage* 2014;48:215-30.
- xxxv. Gaertner J, Eychmueller S, Leyhe T, Bueche D, Savaskan E, Schlögl M. Benzodiazepines and/or neuroleptics for the treatment of delirium in palliative care? - a critical appraisal of recent randomized controlled trials. *Ann Palliat Med* 2019;8:504-15.