Considerations for Dysphagia Management in Neuromuscular Disease

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What is Neuromuscular Disease (NMD)?

Diseases of the:

i)

ii)

iii)

- Hereditary or acquired
- Muscle weakness without impact on sensory functions
Dysphagia in NMD

- Recognised complication
- Prevalence of up to 65-86% in certain subtypes (Cox et al., 2009; Needham and Mastaglia, 2007; Oh et al., 2007; Williams et al., 2003)
- Suspect underestimation
- Strongly linked with malnutrition (Tilton et al., 1998)
- Dysphagia intervention & management increasingly critical as life expectancy increases
- Respiratory disease & respiratory infection leading cause of death
What makes these patients unique?
Approx. 1/3 of DMD have additional intellectual impairment (Bresolin et al., 1994)

Declining fronto-temporal cognitive impairment in DM$_1$ and DM$_2$ (Sansone et al., 2007; Modoni et al., 2008)

Variable prevalence in congenital muscular dystrophies (Messina et al., 2010)
Multi-system disease

- Motor Neurone Disease (MND)
- Myaesthenia Gravis (MG)
- Spinal Muscular Atrophy (SMA)
- Duchenne Muscular Dystrophy (DMD)
- Myotonic Dystrophy (DM1)
- Inflammatory Myopathies (PM & DM)
- Congenital myopathies (CM)
Early dysphagia presentation

Oculopharyngeal muscular dystrophy (OPMD) (Bumm et al 2009; Werling et al., 2015)

Inclusion body myositis (IBM) (Verma et al., 1991)

Diagnostics:
Where are these patients?
Risk factors
Dependence for Feeding

- Upper limb weakness
- Early support required in DMD and SMA
- Independence maintained with adaptive equipment
- Risk factor for aspiration in other patient populations
  (Siebens et al., 1986; Langmore et al., 2002; Marik et al, 2001;)

Activity Levels

- Reduced
- Difficult to isolate
Posture

- Scoliosis common - DMD & SMA (Cambridge and Drennan, 1987; Merlini et al., 1989)
- Poor sitting balance, severely altered posture, restriction on lungs, pain
- Some eat and drink lying down

http://www.mehtaspine.co.uk/scoliosis.php
Respiratory Decline in NMD

The respiratory system:
- Conducting airways
- Gas-exchange airways
- Respiratory muscle pump
- CNS & PNS
- Pulmonary vasculature
Respiratory Assessments

- Overnight sleep studies
- Blood gases (ABGs or CBGs)
- Lung function tests (LFTs)
- Symptom presentation
Non-invasive ventilation

Images from:
What’s the relevance?

• Breath-swallow patterns (Terzi et al., 2007)
• Lung vulnerability
• Effect of hypercapnia on swallow performance?
• Low functional capacity (Gross et al., 2003)
• Poor cough (Bianchi et al., 2012)
Ventilation

• No current research examining direct or indirect impact of NIV on the swallow
• Comparison with small studies on invasive ventilation via tracheostomy
• NIV contraindications:
  – Unable to protect airway from aspiration
  – Poor airway patency
Cough Clearance

- Weak cough
- Physiotherapy provide cough clearance techniques
- Increased risk of developing pneumonia as unable to successfully clear the airway independently (Finder, 2010)
Peak cough flow

- Integrated into the bedside dysphagia assessment
- Normal range:
- Required to clear secretions/debris:
- Prophylactic physio treatment:
Cognition

- Symptom report – dependence on health care professionals to ‘ask the right questions’
- Apathy or indifference
- Compliance to advice
Gastroenterology

- Gastro-oesophageal reflux
- Delayed gastric emptying
- Poor motility
- Constipation
- Bloating/swelling
Dysphagia in NMD

What does it look like?

... and can you spot it from the end of the bed?
Case Example

- 17 year old boy with DMD
- Denied any history of dysphagic symptoms
- No clinical history to suggest dysphagia (one winter chest infection 12 months prior to appointment but nothing since)
- Over-weight, high BMI
- No dysarthria, no dysphonia, no drooling
Case Example

- Acute admission to NIV unit with pneumonia
- Continued to deny dysphagic symptoms
- Food debris identified in cough assist by NIV physiotherapist
- Referred to SLT during acute admission
- Subtle clinical indicators in bedside assessment which took a long time to provoke
- Chest markers linked with trace, chronic aspiration
Hurdles

• Oral antibiotics...
• Acute on chronic respiratory failure – 24 hour NIV
• First chest infection in 12 months
• No robust data/evidence to guide management
• “I’ll only have a PEG if you can show me food going down the wrong way...”
• FVC of < 50%
How accurate is patient report?

- Anecdotal evidence of sensory deficit
- Desensitisation over time?
- Impact of NIV/reflux?
How useful is videofluoroscopy?

- The jury is out
- Silent aspiration
- Silent residues
- Management strategies
Is it ever ‘too late’ for PEG placement?

• NCEPOD report (2004) – MDT decision
• Increased risk of complications as VC falls
• Spinal deformity & anatomical changes
• RIG contraindicated in low BMI or rapid weight loss
Is PEG is the right thing?

- Outcomes & best practice documented in MND (Forbes et al., 2004; Anderson et al., 2007)
- When? How?
- Lack of longitudinal data
Psychological Impact

Grieving

Fear & anxiety
Tips for assessment...

• Take a precise case-history – dig deep!
• Don’t rely solely on patient report
• Ideally make observations over a mealtime
• Not uncommon for foods to be worse than drinks
• Look beyond just ‘the swallow’ - appraise additional risk factors for aspiration
• Low threshold for concern
• Get in early and build rapport
• Be honest
• Educate the MDT
• Don’t rush decision-making
• Pool resources
• Be suspicious... and don’t give up!
• Keep the patient at the centre
References


Useful Reading & Resources

Respiratory Care:
http://erj.ersjournals.com/content/erj/34/2/444.full.pdf
http://thorax.bmj.com/content/67/Suppl_1/i1.full.pdf+html

MDT Care:
Duchenne
SMA
MND
http://www.nice.org.uk/guidance/cg105/evidence/full-guidance-134707501
Myotonic
http://ac.els-cdn.com/S096089661000595X/1-s2.0-S096089661000595X-main.pdf?_tid=45327004-7daa-11e5-bdb7-00000acab361&acdnat=1446060776_ff6d802d24d01413729a8e5faec1a1a9
MND Association
http://www.mndassociation.org/

Muscular Dystrophy Campaign
http://www.musculardystrophyuk.org/

Myaesthenia Gravis Association
http://www.myaware.org/

SMA Trust
http://www.smatrust.org/

Action Duchenne
http://www.actionduchenne.org/