Respiratory issues in children with neuromuscular disease

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Overview

Impact of neuromuscular disease on respiratory health

Respiratory assessment

Therapies

Shared decision making
NMD & respiratory health

Major impact on respiratory health
Respiratory failure commonest cause of death
Can have similar impact to other neurological problems
  - impaired defense
  - airway obstruction
  - restrictive physiology
  - respiratory failure

Key is recognising disease trajectory – onset, severity & progression
British Thoracic Society guideline for respiratory management of children with neuromuscular weakness

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Respiratory problems

Upper airway

Ventilation

Infection
Upper airway

Weak inspiratory, oropharyngeal & expiratory muscles leads to poor cough
Reduced airway secretion clearance
  Increased respiratory infection
Reduced gag & swallow reflexes
Abnormal swallow
  Increased risk of aspiration
Airway tone problems
  Obstructive sleep apnoea
Normal ventilatory balance

Ventilatory drive

Respiratory load

Respiratory muscle capacity

airway
lung
thoracic mechanics
Ventilatory imbalance

- **↑ Ventilatory drive**
- **↓ Respiratory muscle capacity**
- **↑ Respiratory load**

**Alveolar hypoventilation**
- **↓ PaO₂ and ↑ PaCO₂**

**Secretions**
- **Pneumonia**
- **Atelectasis**
Ventilation

Ventilation imbalance worse during sleep
Particularly REM sleep
Can be exacerbated by OSA
Symptoms of nocturnal hypoventilation
  Tiredness on waking
  Morning headache
  Hypersomnia
  Poor concentration
decrease in spirometry

daytime hypoventilation

nocturnal hypoventilation

exacerbation

respiratory failure

Progression

CRISIS
Infections

Increased risk from
  aspiration
  poor secretion clearance
  structural lung damage

Decreased respiratory reserve
Assessment

Clinical assessment at each consultation
Formal assessment best by multi-disciplinary team
  - history
  - examination
  - oximetry
  - non-invasive tests
Respiratory function tests

Spirometry

characteristic restrictive defect
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Respiratory function tests

Spirometry

characteristic restrictive defect

Vital capacity may be a predictor for susceptibility to e.g. infection, hypoventilation

~<60% predicted
Respiratory function tests

Can measure maximal static pressures (MIP & MEP)

Sniff nasal inspiratory pressure (SNIP) easier (inspiratory muscle strength)

May not add much compared to VC
Respiratory function tests

Cough peak flow
(expiratory muscle strength)

Adult CPF >400 lpm
<160 lpm (in >12yr old) reduced clearance

Useful assessment particularly in >12 yo
Sleep assessment

Typical indications: infants with weakness symptoms

At least annually VC < 60%
non-ambulatory

Oximetry (may be sufficient if normal)
Oxycapnography (end-tidal or TC CO2)
Polysomnography (cardio-respiratory or full PSG)
Scoliosis

Common
Contributes to increased respiratory load

MDT assessment prior to surgery important
May need to start/habituate to NIV pre-op
Effective airway clearance essential
NIV can facilitate extubation
Swallow assessment

Clinical assessment
Video fluoroscopy gold standard

Optimum nutrition important
Consideration of gastrostomy
May be balance between risk of aspiration and pleasure from eating
Physiotherapy

Crucial positioning
suctioning
manual physio
(assisted) breath stacking
manual assisted cough
MI-E
(mechanical insufflation/exsufflation)
(cough assist devices)
Treatment

Funding and community physio support can be difficult

Appropriate interventions can reduce admissions

Nebulised saline / hypertonic saline may help

May have gastro-oesophageal reflux

Early treatment with intermittent antibiotics

Prophylactic antibiotics eg azithromycin may help

Appropriate immunisations
Non-invasive ventilation

Consideration based on

disease trajectory
symptoms
respiratory function
sleep assessment
exacerbations
patient & family preferences

↓ symptoms   ↓ admissions   ↑ survival
DMD, Northern Region, UK  Eagle, Neuromusc Dis 2002;12:926

- Ventilated (n=24)
- Died post 1990 (n=33)
- Died in 1980's (n=68)
- Died in 1970's (n=49)
- Died in 1960's (n=9)

NIV
Not ventilated
14 yo
Duchenne muscular dystrophy
FVC 0.7 L (20%)
Asymptomatic, overseas holidays
14.5 yo
Symptomatic – grumpy and tired in mornings

Started NIV
Tracheostomy

NIV is mode of choice for most children

When consider tracheostomy?
- Severe bulbar dysfunction with aspiration
- Failure to extubate
- Ventilation >16 hours per day
- NIV failed

Can have advantages BUT

Risks, adverse effects & critical decision point
Quality of life & palliative care

Palliative care can be delivered at any point (not just at the end of life)
Involvement with team early useful
Parallel planning effective – not opposing strategies
Discussions around preferences around end of life & periods of instability – advance care planning
Issues around LTV & NMD

May be uncertainty about diagnosis & future
Balance of prolonging survival vs risk of diminishing quality of life
How do we assess quality of life?
Balancing benefits vs burden
Resources are limited
Opinions vary; professionals may have biases
Practices & decision making processes may be different and be inconsistent
Case J

Admitted 2 weeks of age
Unexplained episodes of desaturation
Dependence on high flow therapy
Respiratory failure – ventilated 5 weeks of age
Failure to extubate
Case J

Admitted 2 weeks of age
Unexplained episodes of desaturation
Dependence on high flow therapy
Respiratory failure – ventilated 5 weeks of age
Failure to extubate
Abnormal diaphragm movement on USS
Discussion around neuromuscular conditions
Diagnosis of SMARD at 8 weeks of age
Case J

Discussion with parents around diagnosis
Early presentation – likely severe SMARD

Information gathering for family
Counsellled would need 24/24 ventilation &
would have progressive generalised weakness

Frequent meetings PICU/respiratory
Parallel planning with palliative care
Case J

Worsening stability on invasive ventilation
Counselled against tracheostomy & LTV
Case J

Worsening stability on invasive ventilation
Counselled against tracheostomy & LTV

Parents agreed not to pursue LTV

Decided to have elective extubation in PICU
Died 16 weeks of age
Key questions

Who should be making the decisions?

And on what basis?
Legal basis for making decisions

Paramount consideration of the best interests of the child

Statutory welfare checklist

- ascertainable wishes of the YP
- needs
  - capability of parents or others to meet needs

Balancing exercise of gains vs losses

Favours agreement of a child’s best interests
Who is best placed to decide?

We might have knowledge but does that inform the “best” decision?

Professionals can under-value the quality of disabled lives

Parents could be impacted by their emotion – might underestimate hopelessness

“Uncertain locus of moral authority”

Encouraged to have “shared decision-making”
Uncertainty

There is often uncertainty
At best we can only make judgments

We likely need to be more open about presence
of uncertainty and communicate that to families
Shared decision making

Still needs to serve child’s best interests
Effective communication between stakeholders essential
Viewpoints supported & heard
Curative and palliative care shouldn't be seen as opposing strategies
Framework for decision making

Focuses emphasis on **process of deliberation** rather than necessarily the product

Good early & honest communication of facts
Diagnosis is key, but acknowledging uncertainty
Lead MDT, appropriately frequent meetings
Respect for all views
Efforts to understand quality of life within context of family
Parents not to feel they alone carry burden of decision making
Parallel planning
Summary

Impact of neuromuscular disease on respiratory health

Respiratory assessment

Therapies

Shared decision making