Respiratory Management:

Duchenne Muscular Dystrophy

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Respiratory muscle weakness starts about 10 years BUT only becomes a problem when wheelchair mobile
All the following deteriorate together - slowly over time

• **Respiratory muscle weakness**
  - Hypoventilation – under breathing
  - Reduced cough

• **Slow swallowing**
  - Risk of aspiration

• **Scoliosis (curvature of spine)**
  - Causes further restriction on breathing

Slowed by steroids
Breathing - ventilation

• To take in Oxygen ($O_2$)

• To remove Carbon dioxide ($CO_2$)
As muscle weakness increases

- Reduced breathing – reduced chest wall expansion – reduced ventilation

- If not breathing adequately then:
  - reduced Oxygen (O2) in blood
  - Increased Carbon Dioxide in blood

- Comes on gradually: brain ‘sensors’ reset
  - Not like you and me holding our breath!
Respiratory – ventilation failure comes on gradually

Initially:

- **Under or hypo-ventilation occurs at night (asleep):** headache, tired, unduly sleepy by day

- **Reduced cough strength** – when ‘head cold’ and need to clear phlegm from the upper airways: feeling phlegm in windpipe, chest infections

- Cough – 1\(^{st}\) need a deep inspiration
  - need good respiratory muscle power
Mobilization

1. Inspiration
Mobilization

2. Expiration
As time goes on – respiratory muscle weakness increases

• Respiratory failure increases / encroaches into the day

• Complete inability to cough

• Swallowing – increasing risk ‘aspiration’ (food goes into windpipe during swallowing)

Key features
  – Prolonged meal times
  – Fatigue with oral feeds
  – Cough/choking with swallows
The ‘twist’ - scoliosis
Scoliosis in Duchenne MD

Impact of adolescent growth spurt
How can we monitor and treat reduced breathing

Inadequate cough

‘hypoventilation’
Peak expiratory cough flow (> 250 L)

Potential problem
PCF < 150

History: inability to cough adequately
Once inability to cough adequately
Peak Expiratory Cough Flow < 100

Need ‘cough assist’ techniques
• Standard chest physiotherapy
• Physio – breath stacking
• Cough augmentation physio’
• Cough assist devices
Vital Capacity

FVC

FVC < 1.2 L ---- worry about hypoventilation

FVC < 1.0 L ----- should need ventilation support
Night time hypoventilation

- Forced Vital Capacity < 1.2 Litres
- Symptoms:
  - Morning headache, tired, sleepy by day
  - Reduced performance at school
- Should be having annual Overnight Oximetry
  - Measuring Oxygen levels overnight in home
Monitoring Carbon Dioxide
(Transcutaneous CO2)

• Start doing this when FVC < 1.2 Litres
• Oximetry is abnormal

• Usually needs overnight hospital study
  CO2 should be 4 - 6/7
If hypoventilation ...

- Tc CO2

- Typically at night

- Need to give some **support** to breathing
  - this is NOT taking over breathing – just adding extra to each normal breath.
Non-invasive ventilation (NIV)

• **BiPAP**: Bilevel positive airway pressure (BiPAP)

• ‘bi’ = 2 levels of pressure
  – inspiratory pressure eg 12 – added to normal breath
  – End expiratory pressure eg 5 (so that the small airways don’t collapse down)

• Makes a vast difference to day time function
Pulmonary Care
Dysfunctional swallowing

• Speech & Language Therapist
  – Feeding history
  – “DOM” – directly observed meal

• Videofluoroscopy swallow study
  – Not just diagnostic of potential aspiration
  – Opportunity to evaluate strategies using ‘positioning’ and ‘thickening / consistencies’

• Sometimes a decision needs made regarding N/G or peg-tube feeding
Pulmonary care

• Prevent infections vaccinations:
  — Annual Flu’ jab
  — pneumococcus,
  — RSV
From the internet
“My son, Roy, with DMD is 32 and has used the night time v-pap with nasal mask and daytime volume vent with mouth interface (“sip-vent”) for over 10 years and has thus avoided a trach.”

Tracheostomy
RESEARCH
When we start
  1. Cough assist
  2. BIPAP

We don’t know how to optimise the lung expansion R. v L.

If scoliosis –
  one side doesn’t expand,
  more likely to ‘collapse’
  and be infected
**We need a way to measure Right v Left lung expansion**

In Spinal Muscular Atrophy
  1. diaphragm relatively spared
  2. ‘paradoxical breathing’

We need to be able to measure CHEST expansion
  versus Abdominal movement
Structured Light Plethysmography

- Novel, non-invasive device
- Grid projection onto anterior chest and abdominal wall
- Changes in grid movement captured

- Corresponding virtual 3-D chest wall surface created
- Allows interpretation of respiratory volume and regional chest wall expansion
Figure 1. Working principle of SLP.
Patient with mild Scoliosis -- starting BiPAP.
The SLP measurements before & after BiPAP
Starting low pressures used (10/5) BiPAP.

**NO need to put pressures higher**

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<thead>
<tr>
<th></th>
<th>Room air:</th>
<th>On BiPAP:</th>
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<tbody>
<tr>
<td>Right lung</td>
<td>55</td>
<td>50</td>
</tr>
<tr>
<td>Left lung</td>
<td>45</td>
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ANY QUESTIONS –