

Neuromuscular disorders (PIP)

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Key Documents Owner:	Dr Gallagher	Consultant Paediatrician
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The following guidance is taken from the Partners In Paediatrics (PIP)

Neuromuscular disorders 2018-20

NEUROMUSCULAR DISORDERS

ON ADMISSION

- Ask parents if they have a copy of a care plan
- Inform child's long-term consultant

CLINICAL HISTORY

- Adequacy of cough and swallowing
- Previous sleep difficulties, wakefulness at night (nocturnal hypoventilation)
- Difficulty waking in morning, early morning headache (nocturnal hypoventilation)
- Poor appetite, weight loss (chronic respiratory failure)
- Learning or behavioural problems, school **absence** (chronic respiratory failure)
- Palpitations, breathlessness, chest pain (cardiomyopathy)
- Muscle cramps, skeletal pain, back pain (for fractures)
- Abdominal pain, distension, melaena (GI perforation)

ASSESSMENT

- May not show overt signs of respiratory distress such as tachypnoea, recessions and use of accessory muscles even in respiratory failure
- Assess adequacy of chest wall excursion and cough
- Look for pallor, tachycardia, signs of circulatory compromise
- Assess for abdominal signs (GI bleed, perforation, gastritis)
- Measure:
 - SpO₂ in air
 - CO₂ by blood gas, transcutaneous CO₂ or end-tidal CO₂, especially if on oxygen
 - spirometry: FVC most useful if previous readings available
- ECG
- Blood gas for cardiac status
- CXR: clinical signs can fail to detect collapse/consolidation/cardiomegaly
- Consider skeletal/spinal X-rays for possible fractures

Medical problems commonly found in children with myopathy

- Respiratory failure (hypoxaemia and hypercapnia) without signs of respiratory distress. Susceptibility to respiratory failure due to:
 - muscle weakness (upper airway, intercostals, diaphragm)
 - scoliosis
 - poor secretion clearance
 - aspiration, chest infections
 - sleep disordered breathing
 - cardiac failure
- Lower respiratory infection, aspiration pneumonitis
- Cardiomyopathy and cardiac decompensation
- Gastro-oesophageal reflux, gastritis and gastric ulceration (especially if on corticosteroids)
- Adrenal insufficiency (if on corticosteroids)
- Fractures, especially vertebral, if on long-term corticosteroids
- Malignant hyperthermia following anaesthesia in certain muscular dystrophies and myopathies

MANAGEMENT

- If unwell, on **long-term corticosteroids**, double usual daily dose of steroids for 2–3 days. If unable to tolerate oral steroids, see **Steroid dependence** guideline

Respiratory failure

- Prescribe and carefully titrate administration of oxygen by mask/nasal cannulae to achieve SpO₂ between 94–98%. Monitor CO₂ and respiratory effort as risk of rising CO₂ and respiratory failure (despite normal oxygen saturations) if hypoxic respiratory drive overcome by oxygen therapy
- High-flow high-humidity air or oxygen (e.g. Optiflow™): monitor CO₂
- Mask ventilation (bi-level positive airway pressure, BIPAP)
- Chest physiotherapy and postural drainage
- Use insufflator-exsufflator (e.g. Cough Assist) if patient has one
- Suction
- if copious loose secretions, use glycopyrronium given as oral solution (Sialanar) or IV solution (200 microgram/mL) given orally, via PEG or IV
- if thick tenacious secretions use nebulised sodium chloride 0.9%/sodium chloride 3%, or nebulised acetylcysteine
- Antibiotics
- obtain cough swab or sputum specimen, ideally before starting treatment
- check previous culture results
- choice same as for community acquired pneumonia
- if bronchiectasis use broad spectrum for 14 days to cover pseudomonas (discuss with senior)
- if not improving on first line antibiotics add macrolide for atypical pneumonia
- Consult senior to discuss need for ITU care, escalation of respiratory support

Cardiac failure

- Fluid restriction
- Diuretics
- Oxygen and respiratory support
- Cardiology consultation

GI tract bleed: prevention and treatment

- Nil-by-mouth and IV fluids
- Ranitidine (omeprazole if severe reflux)
- Senior advice

Fractures

- Analgesia
- Orthopaedic consultation
- Check calcium and vitamin D
- Discuss with metabolic bone expert about IV bisphosphonates for vertebral fractures

Malignant hyperthermia

<i>Malignant hyperthermia is a medical emergency</i>

- Occurs following general anaesthesia and may be first presentation of a neuromuscular disorder
- Check creatine kinase, calcium, renal function, urine output and for myoglobinuria: dialysis may be needed
- In addition to temperature control and general life support measures, use IV dantrolene to control excessive muscle contraction
- Obtain senior anaesthetic advice and liaise with PICU