Expecting the unexpected: Pre-operative protocol created after a 17-year-old girl with MPS IV (Morquio syndrome) and cystic fibrosis died from an intra-operative spinal injury

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Background:

• A 17-year-old girl, from non-consanguineous parents
• Dual diagnoses of MPS IVA (Morquio syndrome) and cystic fibrosis
• Required semi-urgent laparoscopic cholecystectomy (unrelated to her MPS IVA)
• Duration of procedure was much longer than expected at 229 minutes
• Post operative complications: diaphragmatic weakness, urinary incontinence, significant lower limb weakness and abnormal sensation
• An MRI showed a thoracic spinal cord injury suggestive of an infarct
• The patient and family elected to return to theatre for a decompressive laminectomy and spinal fusion
• Her condition rapidly deteriorated and she succumbed to her intra-operative injuries 17 days after the first operative procedure

Review process:

• A comprehensive post-mortem Human Error and Patient Safety Training Program (HEAPS) review occurred
• The review recommendations were to develop an evidenced based tool for clinicians, patients and families to inform them of the perioperative risks and to ensure that all staff involved in the management of children and adults with MPS or other skeletal dysplasias understood the risks prior to booking any surgical procedure

Conclusion: A comprehensive work instruction was developed by the Director of Metabolic Medicine in consultation with metabolic physicians, departments of Anaesthetics, ENT Otolaryngology and Head & Neck, Respiratory Medicine, Rehabilitation and the Patient Quality and Safety Committee and endorsed by the hospital executive, to guide staff in the preparation and planning for surgical procedures to ensure all potential perioperative risks are identified and appropriate preventive management plans arranged prior to proceeding to a surgical booking.
Work Instruction

Mucopolysaccharidoses (MPS) airway management - Pre-operative planning pathway

Purpose

The purpose of this document is to provide instruction for Children’s Health Queensland (CHO) staff who in their daily work are involved in assessing, booking or performing procedures that require children with a mucopolysaccharidosis disorder to undergo a general anaesthetic to minimize undesirable clinical outcomes.

Scope

This work instruction applies to all clinical staff who are involved in managing this patient children with MPS disorders.

Instruction

Children with a diagnosis of a mucopolysaccharidosis (MPS) disorder pose significant risk for airway complication and spinal injury during procedures that require a general anaesthetic.

A thorough airway assessment should be undertaken prior to going to theatre. In doing so, consider the following:

Assessment

• At time of referral, notify the Metabolic Team of the planned procedure
• Notify all other teams involved in the patient’s care (e.g. anaesthetic, surgical, respiratory, ENT, cardiac (if needed))
• Assess as a multi-disciplinary team the risks and benefits of the procedure for the child/young person then discuss the combined clinical opinion with the young person and their parent/guardian

Anticipation

• Plan for and anticipate problems
  o Discuss with the ENT or Respiratory Team regarding the need for one of their team to be present on induction

Anticipate

• A grade IV airway
• High risk for spinal injury
• Increased risk of bleeding for MPSI, II, III
• Severe behaviour disorder for the neurological forms of MPS II, III
- Airway complications – tortuous trachea +/- glycosaminoglycan (GAG) deposits in the airway
- Spinal cord injury – especially in spinal surgery but also known to occur in general surgery
- The need for at least 1 night’s stay post procedure

Consideration
- Consider best option for OT list: e.g. day of week, availability of ENT surgeon or respiratory physician (unless in an emergency) where possible avoid an after-hours or weekend list
- Consider the child/young person’s usual sleep position
- Consider the need for a PICU bed depending on the type of surgery
- Consider the need for neurophysiological monitoring (discuss with the surgeon and metabolic team)
- Develop a management plan for each possible complication

Communication
- If booking a procedure for a child with an MPS disorder, notify
  - The metabolic, ENT, respiratory, anaesthetic Teams and cardiac team if relevant
  - Using the Mucopolysaccharidoses (MPS) Anaesthetic Consent form, the metabolic team will fully inform the young person, parent/guardian of the risks of the procedure (pertinent to the MPS disorder)
  - The surgical/orthopaedic team will undertake the process of informed consent with the young person and parent/guardian of the risks of the procedure from a surgical perspective
  - After an anaesthetic review appointment, the Anaesthetic team will undertake the process of informed consent with the young person and parent/guardian of the risks of the procedure from an anaesthetic perspective by using the Pre-Anaesthetic Checklist for Mucopolysaccharidoses/Lysosomal Storage Disorders.
- Once the metabolic team are aware of an OT booking that involves a child with an MPS diagnosis, the metabolic team will
  - Email the OCH-Anesthesia@health.qld.gov.au with a copy email to the Director of Anaesthetics, Director of ENT, Director of Respiratory Medicine and the Director of Metabolic Medicine as well as a copy to the treating Respiratory, cardiac and ENT physician (if the child has one)
    - The email will include
      - Child’s name and medical unit record number
      - Diagnosis
      - Date of procedure
      - Planned procedure and surgical/orthopaedic team

Equipment

It is mandatory that access to a flexible bronchoscope is available to access within QCH throughout the planned procedure should this be clinically required.

Supporting documents

Forms and Templates
- Pre-Anaesthetic Checklist for Mucopolysaccharidoses/Lysosomal Storage Disorders
- Mucopolysaccharidoses (MPS) Anaesthetic Consent
## Pre-Anaesthetic Checklist for Mucopolysaccharidoses/Lysosomal Storage Disorders

**Home Team:**

**SMO/NP:**

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### Team emergency contact details:

<table>
<thead>
<tr>
<th>Assessment</th>
<th>Required</th>
<th>Date performed</th>
<th>Signature</th>
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<tbody>
<tr>
<td>Anaesthetic assessment</td>
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<td>Venous access</td>
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<td>PIV  PICC  PORT</td>
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<td>Pre-Op bloods</td>
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<td>Bloods needed under GA</td>
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<td>• Respiratory assessment</td>
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<td>• Respiratory function tests</td>
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<td>• (in the last 6/12)</td>
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<td>• Sleep study (if CLINICALLY indicated)</td>
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<td>ENT assessment (in the last 6/12)</td>
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<td>• Cardiology assessment</td>
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<td>• ECHO/ECG (in the last 6/12 if CLINICALLY indicated)</td>
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<td>• Imaging</td>
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<td>• Flexion/extension spinal X-rays</td>
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<td>• 3D CT Trachea and Chest</td>
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<td>(in the last 6/12 if CLINICALLY indicated)</td>
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<td>• MRI Full Spine (in the last 1-2 years)</td>
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<td>Mouth opening measurement</td>
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<td>Severe behaviour disorder</td>
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<td>Psychology/play therapy</td>
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<td>Special fasting requirements</td>
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<td>Overnight pre-admission needed</td>
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<td>ICU/HDU bed required</td>
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**Usual sleep position:**

**Other comments/concerns:**

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**Have you notified the metabolic team of planned procedure?**

- **Yes**
- **No**
Authors would like to acknowledge the Willink Biochemical Genetics Unit team at St Mary’s Hospital, Manchester for the information provided in the development of these tools.