BIMDG

British Inherited Metabolic Disease Group

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# ANAESTHESIA IN PATIENTS WITH MUCOPOLYSACCHARIDOSES (MPS)

**WARNING: General anaesthesia in many patients with mucopolysaccharidoses is potentially dangerous. It should be planned in consultation with, and administered by, an anaesthetist experienced in managing difficult paediatric airways and in centres with access to an appropriate ICU as admission to PICU may be necessary post-operatively.**

For further advice please contact one of the paediatric metabolic units that are designated centres for management of MPS: Royal Manchester Children's Hospital; Birmingham Children's Hospital; Great Ormond Street Hospital, London;

1. Deposition of mucopolysaccharide leads to involvement of many organ systems. In addition to routine pre-operative investigations, special attention should be given to the cardio-respiratory system, even in the absence of signs and symptoms. Pre-operatively all patients should have:
   ECG
   Echocardiogram
   Chest X-ray
   Lung function tests are necessary for those with kyphoscoliosis.

2. Upper airway problems should be anticipated and assessed pre-operatively. A history of sleep apnoea should be sought, and radiographic assessment of the upper airway considered. Copious secretions are troublesome and pre-medication with an anti-sialagogue may be helpful.

3. Obstruction following induction of anaesthesia may occur and it may not always be possible to achieve a clear airway.

4. Intubation may be very difficult for several reasons. These include a relatively large head and short immobile neck, macroglossia, sometimes accentuated by micrognathia, and mandibular abnormalities which limit mouth opening. Infiltration of soft tissues in the upper airway adds to the problems. The size of the tracheal tube required is commonly smaller than estimated for the age of the child, possibly due to mucopolysaccharide infiltration of the airway.

5. Patients are at risk of atlantoaxial subluxation secondary to hypoplasia of the odontoid peg. This is a particular problem in Morquio disease. In addition, the cervical spinal canal may be narrowed, and this may lead to progressive myelopathy. Excessive flexion or extension of the neck during tracheal intubation and anaesthesia may therefore lead to cervical cord damage. Immobilisation of the neck by a halo brace or plaster jacket may offer some protection.

6. If hepatosplenomegaly is marked, the possibility of regurgitation should be considered.

7. Central nervous system involvement leads to difficulty in handling these children, especially in Sanfilippo disease when they may be quite aggressive.

8. Skeletal deformities may cause difficulty in positioning for surgery, and in the event of cardio-respiratory arrest, may impede resuscitation.

9. Post-operative respiratory problems may occur, including upper airway obstruction, post-obstructive pulmonary oedema and chest infections, due to thick secretions. Peri-operative physiotherapy may be required. In selected, high risk patients the provision of HDU/ ICU cover post-operatively must be sought.

10. Local and regional anaesthetic techniques are usually unsuitable in these patients because of poor cooperation. However, they may be possible in older patients with normal or near normal intelligence.

11. Prolonged bleeding times may be seen in some patients (e.g. those with Sanfilippo disease) and care must be taken especially with adenotonsillectomies.

12. General anaesthesia is very risky in these patients and every effort must be made to combine different procedures and examinations under one general anaesthetic.

In spite of all these difficulties, or rather because of them, it should be emphasised that a general anaesthetic may be safer than sedation, because it is not possible to control the airway of a sedated child. Most procedures in these patients are therefore best carried out under general anaesthesia.

## CONCLUSION

Close liaison with the anaesthetic department is essential when any procedure is contemplated in one of these patients. The benefits of the procedure/surgery should always be weighed against the risk of anaesthesia.

## CHECKLIST

The following points must be checked while clerking in the patient.

1. HISTORY
   - Previous problems with anaesthetics
   - Obstructive sleep apnoea (may not be obvious- ?difficulty in sleeping ? night terrors).
   - Vertigo, transient paresis/paralysis esp. after falling (cervical instability).

2. EXAMINATION
   - Excessive nasal/pharyngeal secretions
   - Blood pressure
   - Kyphoscoliosis (may cause problems with positioning).
   - Evidence of spinal cord compression.

3. PRE-ANAESTHETIC INVESTIGATIONS

Check to see if the following have been done:

- Chest Xray
- Echocardiogram/ECG
- Lung function tests.
- Sleep studies
- Xrays of the cervical spine to look for instability

A chest Xray and echocardiogram or ECG are essential. If an echocardiogram has not been performed within the last 6 months, then it must be performed prior to anaesthesia. The others are not essential, but if they have been done, and the result was abnormal, then the anaesthetist must be notified.

4. LIAISON WITH ANAESTHETIST

The anaesthetist should be notified of the following

- Excessive secretions; ? need for sialogogues
- Obstructive sleep apnoea, either clinical or recorded
- Previous anaesthetic problems
- CXR shadowing (Xrays should be available)
- Abnormal results of any other relevant investigations (lung function, cervical instability on plain films)
- Evidence of spinal cord compression (either clinical or MRI)

**IMPORTANT: There may have been significant disease progression since the last time the patient was seen in the clinic. This is particularly true of MRI scans. Therefore a patient who was fit when last seen may no longer be fit. Please ensure, therefore, that all the points on the checklist have been 'ticked'.**


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