INFORMATION for Anaesthetists

This leaflet outlines:

- **the symptoms of MPS II, and the clinical consequences relevant to anaesthesia**
- **considerations for MPS II patients before, during and after surgery**

This leaflet is a summary of the published information as of February 2018, is not exhaustive and does not constitute any recommendations. It is the responsibility of the anaesthetist and the multidisciplinary team to determine the optimal management plan for the individual patient.

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The information in this leaflet is intended for healthcare professionals only.

This information is intended for an international audience outside of the USA and has been produced by Takeda. The leaflet has been developed in accordance with industry and legal standards to provide information for healthcare professionals about Hunter syndrome health topics. Takeda makes every reasonable effort to include accurate and current information. However, the information provided in this leaflet is not exhaustive.

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MPS II

A combination of common childhood complaints in your patient could indicate Hunter syndrome (MPS II), a rare genetic disorder that predominantly affects males. MPS II is caused by a mutation that stops the lysosomal enzyme iduronate-2-sulfatase (I2S) from being produced. I2S normally breaks down molecules called glycosaminoglycans (GAGs); without I2S, GAGs accumulate, causing progressive, multisystemic disease, leading to early mortality.[1]

Symptoms of MPS II include otitis media, abdominal hernia, nasal obstructions, and enlarged tonsils/adenoids and tongue.[2] Although these symptoms are common among children, in MPS II patients these symptoms occur early and in combination, and may be refractory to treatment.[3] This means that individual MPS II patients often have multiple surgical interventions and associated anaesthesia procedures. For example, 40% of MPS II patients have had more than one hernia repair.[3]

Due to the nature of MPS II, anaesthesia and surgical procedures are best performed with careful and specific multidisciplinary management.[2] It is therefore favourable to be able to diagnose MPS II in patients early, so that appropriate management may be initiated before additional procedures take place.

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Considerations for anaesthesia

Sedation and general anaesthesia are high-risk procedures for MPS II patients, due to the anatomic abnormalities inherent to the disease, which lead to positioning problems.[4] In 22% of MPS II patients there are difficulties with intubation, and in 4% of patients it is not possible to extubate at all.[3] Symptoms that contribute to these positional difficulties in MPS II patients include chest deformities, an enlarged abdomen, joint contractures, short neck, immobility of the jaw, and airway obstruction caused by an enlarged tongue, throat or trachea.[2,4]

The following summary describes the current opinion in the literature (as of February 2018), however, it is the responsibility of the anaesthetist and the multidisciplinary team to determine the optimal management plan for the individual patient.

Before surgery

Preparation before surgery is key in MPS II. Patients should be assessed by a multidisciplinary team that includes a cardiologist, an ENT specialist, and an anaesthetist, with a full cardiac assessment to evaluate problems such as valve degeneration.[2]

Specifically, to prepare the anaesthetist for the anatomical/positional obstacles that they will encounter, a flexible nasendoscopy, computed tomography scanning of the airway, or a video-recorded bronchoscopy of the airways with a flexible fibre-optic bronchoscope are valuable assessments prior to surgery.[2,4] The severity of a patient's sleep apnoea, assessed with a sleep study or a formal polysomnography, can also indicate if there are respiratory obstructions.[2] To assess the atlantoaxial instability and fragility of the neck, cervical spine flexion/extension is also useful.[2]

Parents should typically be consulted about the possibility of an acute airway obstruction resulting in an urgent need to perform a tracheotomy or cricothyrotomy during either intubation or extubation.[4]

During surgery

It is useful to have an ENT specialist or paediatric pulmonologist available during the induction of anaesthesia and intubation of the patient, to provide support in the event of complications.[4] It is also good practice to plan multiple surgical procedures for a single anaesthesia session, although extending the operation time increases the risks of respiratory complications.[2,4]

Intubating MPS II patients over a flexible bronchoscope allows any abnormal anatomy or dynamics of the airway to be documented, and contributes to the long-term management of the patient.[4] Tracheomalacia in MPS II makes endotracheal intubation problematic.[4] Care may also be necessary to prevent any spinal cord compression through atlantoaxial instability and hyperextension of the neck.[5]

For difficult intubations or for brief procedures, a laryngeal mask airway may provide adequate airway control.[4]

After surgery

After surgery there is still a great risk of airway complications in MPS II patients. Oedema of the larynx and other tissue in these patients can make extubation difficult, if not impossible. Early extubation of these patients can reduce the risk of a post-obstruction pulmonary oedema exacerbating upper-airway obstructions. Using a helium–oxygen breathing mixture at the time of extubation may relieve the upper airway obstruction and improve outcomes, as the reduced density of this air mixture decreases the work of breathing, and increases linear flow rates.[4]

Recovery from anaesthesia can be slow in MPS II patients, and patients may be unable to maintain an airway after extubation, requiring urgent re-intubation or tracheostomy: postprocedure oedema has been reported as late as 27 hours after surgery.[2]

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Summary

MPS II patients have symptoms that typically require frequent surgical and associated anaesthetic procedures, often involving multidisciplinary management from an experienced team.[2,3] To prepare for the anatomical and positional obstacles that anaesthetists will encounter, a flexible nasendoscopy, computed tomography scanning of the airway, or a video-recorded bronchoscopy of the airways with a flexible fibre-optic bronchoscope can be useful.[2,4] It is effective to intubate MPS II patients over a flexible bronchoscope, but care is needed to prevent spinal cord compression through atlantoaxial instability, or hyperextension of the neck.[4,5] After surgery there is a risk of postprocedural oedema, so monitoring is key.[4]

References:

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