INFORMATION for Paediatric Surgeons

This leaflet outlines:

- **the common symptoms of MPS II**
- **information about the surgeries frequently performed on MPS II patients**

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 paediatric surgeon and the multidisciplinary team to determine the optimal management plan for the individual patient.

Visit **Huntersyndrome.info** to learn more

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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INFORMATION for Paediatric Surgeons

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, multi-systemic disease, leading to early mortality.[1]

Early symptoms that should raise suspicion 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]

It is important to be able to diagnose MPS II in patients early, so that appropriate management may be initiated before additional procedures take place.[4]

ACT EARLY

Check the **abdomen** for **hernia**.

+

Check the ear, nose and throat for **otitis media** and **enlarged tonsils** or **adenoids**.

+

Check the rest of the body for **joint stiffness** and **prominent facial features**.

If you suspect Hunter syndrome: REFER TO A SPECIALIST TODAY

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Presentation

The course and presentation of disease for each MPS II patient is unique, but there are common features that are prevalent, due to the abnormal accumulation of GAGs in soft tissues. These features are noticeable from an early age, and include a large head circumference, a broad nose and flared nostrils, prominent brow, large jowl, thick lips, and an enlarged, protruding tongue.[2]

Common clinical symptoms of MPS II include frequent hearing loss due to otitis media or other conductive impairment or sensorineural degeneration, and breathing and chewing/swallowing difficulties due to respiratory obstructions (enlarged tonsils and adenoids, nasal obstructions), skeletal deformities and abdominal distension.[2,4]

Clinical consequences

Tympanostomy, adenoidectomy and tonsillectomy are among the most common surgical procedures performed to relieve symptoms in MPS II patients.[3] For example, fewer than 10% of children in the general population receive tympanostomies, whereas over 51% of MPS II patients have this procedure.[3] Overall, 57% of MPS II patients undergo surgery before a diagnosis of MPS II is made, and 84% of all MPS II patients undergo surgery.[3]

Hearing loss is a concern in MPS II patients, as it can cause behavioural and learning difficulties (in addition to any underlying cognitive impairment). Upper airway obstructions can cause obstructive sleep apnoea in patients, making them fatigued and also contributing to behavioural problems.[4] These obstructions are also a major contributor to mortality in MPS II patients; 46% of deaths in MPS II patients are due to respiratory/airway involvement, so it is important to manage airway obstructions.[5]

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

Considerations for surgery

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

Anaesthesia is a high-risk procedure in MPS II patients, due to intubation difficulties arising from its characteristic anatomical deformities, and the high chance of postprocedural airway oedema.[4] Before surgery it is beneficial for the surgeon to work closely with the anaesthetist to understand the unique anatomical positioning and dynamics of the MPS II patient. It is also 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 good practice to plan multiple surgical procedures for a single anaesthesia session, although extending the operation time increases the risks of respiratory complications. After surgery there is still a great risk of airway complications, so post-procedural monitoring is valuable.[4]

Summary

Common MPS II symptoms include otitis media, abdominal hernia and enlarged tonsils/adenoids; tympanostomy, adenoidectomy and tonsillectomy are among the most frequent surgical procedures performed. Patients often have multiple surgical interventions procedures, and 84% of all MPS II patients undergo surgery at some point.[3]

Preparation before surgery is key: patients should be assessed by a multidisciplinary team that includes a cardiologist, an ENT specialist, and an anaesthetist, to evaluate any anatomical obstacles to anaesthesia or problems such as valve degeneration.[2,4] An ENT specialist or paediatric pulmonologist can provide support during surgery in the event of complications, and it is also good practice to plan multiple surgical procedures for a single anaesthesia session.[4] After surgery there is still a great risk of airway complications MPS II, so monitoring is important.[4]

References:

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