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Maroteaux-Lamy syndrome

McCune-Albright syndrome

orphananesthesia

a project of the German Society
of Anaesthesiology and Intensive Care Medicine

SUPPLEMENT NR. 11 | 2017

OrphanAnesthesia –
ein krankheitsübergreifendes Projekt des Wissenschaftlichen Arbeitskreises Kinderanästhesie der Deutschen Gesellschaft für Anästhesiologie und Intensivmedizin e.V.

Ziel des Projektes ist die Veröffentlichung von Handlungsempfehlungen zur anästhesiologischen Betreuung von Patienten mit seltenen Erkrankungen. Damit will Orphan Anesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

Patienten mit seltenen Erkrankungen benötigen für verschiedene diagnostische oder therapeutische Prozeduren eine anästhesiologische Betreuung, die mit einem erhöhten Risiko für anästhesieassoziierte Komplikationen einhergehen. Weil diese Erkrankungen selten auftreten, können Anästhesisten damit keine Erfahrungen gesammelt haben, so dass für die Planung der Narkose die Einholung weiterer Information unerlässlich ist. Durch vorhandene spezifische Informationen kann die Inzidenz von mit der Narkose assoziierten Komplikationen gesenkt werden. Zur Verfügung stehendes Wissen schafft Sicherheit im Prozess der Patientenversorgung.

Die Handlungsempfehlungen von OrphanAnesthesia sind standardisiert und durchlaufen nach ihrer Erstellung einen Peer-Review-Prozess, an dem ein Anästhesist sowie ein weiterer Krankheitsexperte (z.B. Pädiater oder Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, so dass die Handlungsempfehlungen grundsätzlich in englischer Sprache veröffentlicht werden.

Ab Heft 5/2014 werden im monatlichen Rhythmus je zwei Handlungsempfehlungen als Supplement der A&I unter www.ai-online.info veröffentlicht. Als Bestandteil der A&I sind die Handlungsempfehlungen damit auch zitierfähig. Sonderdrucke können gegen Entgelt bestellt werden.

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OrphanAnesthesia –
a common project of the Scientific Working Group of Paediatric Anaesthesia of the German Society of Anaesthesiology and Intensive Care Medicine

The target of OrphanAnesthesia is the publication of anaesthesia recommendations for patients suffering from rare diseases in order to improve patients' safety. When it comes to the management of patients with rare diseases, there are only sparse evidence-based facts and even far less knowledge in the anaesthetic outcome. OrphanAnesthesia would like to merge this knowledge based on scientific publications and proven experience of specialists making it available for physicians worldwide free of charge.

All OrphanAnesthesia recommendations are standardized and need to pass a peer review process. They are being reviewed by at least one anaesthesiologist and another disease expert (e.g. paediatrician or neurologist) involved in the treatment of this group of patients.

The project OrphanAnesthesia is internationally oriented. Thus all recommendations will be published in English.

Starting with issue 5/2014, we'll publish the OrphanAnesthesia recommenations as a monthly supplement of A&I (Anästhesiologie & Intensivmedizin). Thus they can be accessed and downloaded via www.ai-online.info. As being part of the journal, the recommendations will be quotable. Reprints can be ordered for payment.

A survey of until now in A&I published guidelines can be found on:

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Anaesthesia recommendations for patients suffering from

Maroteaux-Lamy syndrome

Disease name: Maroteaux-Lamy syndrome

ICD 10: E 76.29

Synonyms: Mucopolysaccharidosis Type VI; MPS VI; arylsulfatase B (ARSB) deficiency

Maroteaux-Lamy syndrome is an autosomal recessive disease caused by deficiency of the lysosomal enzyme n-acetylgalactosamine 4-sulfatase (aryl-sulfatase B) which is involved in glycosaminoglycan (GAG) degradation [1,2]. Progressive accumulation of dermatan sulfate in nearly all tissues is believed to provoke the clinical symptoms associated with MPS VI. GAGs are an endotoxin like molecule that incites an inflammatory response via a tumour necrosis factor pathway and promotes apoptotic cell death of chondrocytes.

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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Disease summary

The estimated birth prevalence is 1 in 320,000 live births in Europe. There is no current worldwide incidence rate, and numbers may range according to country or specific ethnic populations studied. There are between 50 and 300 patients in the USA and approximately 1,100 patients in the developed world with MPS VI. Rapidly progressive forms usually present before two years of age with severe dysostosis multiplex and coarse facial features. Without proper treatment patients succumb before the 2nd or 3rd decade. A more slowly progressive (attenuated) form has been described with later onset, clinical symptoms in fewer systems, less pronounced dysostosis multiplex and longer survival [3,4].

Typically, adult height in the severe phenotype is less than 120 cm and dysmorphic appearance includes coarse facial features, frontal bossing, depressed nasal bridge, enlarged tongue and gingival hypertrophy. Other deformities like thoracic deformities (pectus carinatum), scoliosis or kyphosis (gibbus), macrocepahly, hepatosplenomegaly, protruding abdomen, inguinal and umbilical hernias. The characteristic skeletal dysplasia includes short stature, dysostosis multiplex (in x-ray short thickened metacarpals, abnormal vertebral bodies, paddle shaped ribs and short thick clavicles), and degenerative joint disease. Oral, pharyngeal and upper airway obstruction is common. Both obstructive and restrictive respiratory disease is often present. Obstructive disease is related to bronchial narrowing and tracheobronchomalacia whereas restrictive disease is due to the small stiff thoracic cage and abdominal distension combined with kyphosis, scoliosis, and lumbar lordosis [5].

Cardiac involvement is frequent and an important cause of morbidity and mortality. The primary cardiac manifestation of MPS VI is progressive valve degeneration with stenosis and/or incompetence. Azevedo et al. reported mitral valve regurgitation (96%), tricuspid regurgitation (71%) and aortic regurgitation (43%) in 28 patients with MPS VI. Abnormal electrocardiograms (ECGs) occur in ¾ of all patients with sinus tachycardia and right and left axis deviation most common [2]. Heart failure may emerge due to cardiomyopathy, fibroelastosis, valvular involvement, and pulmonary hypertension. Although coronary artery disease has been described only for MPS 1 its presence should be considered in MPS I, II, VI, VII patients.

Intellect is normal but significant learning issues may arise from hearing and visual limitation. Common neurological manifestations include carpal tunnel syndrome, spinal cord or nerve root compression, optic nerve injury, jugular foramen stenosis and communicating hydrocephalus. Spinal cord compression (SCC) is the result of spinal canal stenosis due to small thickened posterior elements, odontoid dysplasia, thickening of the dura, ligamentum flavum and cruciate ligaments, disk bulging or any combination of these. Stenosis can be exacerbated by the presence of flexion extension instability or gibbous deformity. Patients with MPS VI frequently experience myelopathy associated with SCC during childhood. Patients with SCC in rapidly progressive MPS VI require decompression surgery at a median age of 12 years whereas those with slowly progressive disease did not require surgery until 24 years of age [6,7].

Visual impairment is common (40% of MPS VI). Corneal opacification of varying severity (38% severe opacification) is frequently seen as well as refractive errors, glaucoma, retinopathy and optic nerve swelling and ocular hypertension [8].

Intravenous enzyme replacement therapy (ERT) by galsulfase (Naglazyme®) may improve certain somatic symptoms but not alleviate neurological symptoms. The enzyme does not reach poorly vascularised sites such as corneas and joint cartilage [9, 10, 11]. Bone marrow or haematopoietic stem cell transplantation (HSCT) has been used in rare cases to treat MPS VI patients [12, 13, 14, 15, 16, 17, 18, 19].

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Typical surgery

In younger children, the most frequent surgical interventions include adenotonsillectomy, middle ear ventilation tubes and inguinal or umbilical hernias [20]. Tracheostomy for upper airway obstruction may be necessary in advanced stages of the disease. With the advent of ERT the requirement of every week intravenous enzyme administration necessitates central venous access devices (CVADs). Older children present for dental procedures, carpal tunnel surgery and neurosurgery [21,22].

The most common procedures for SCC are laminectomy, laminotomy and open door laminoplasty (expansion of spinal canal). Often a foramen magnum craniectomy is also performed because the compression involves the upper cervical cord at the foramen magnum [23,24,25,26]. Cardiac surgery for valve repair or replacement is more common in the severe form but the literature is a scarce with respect to cardiac surgery interventions in MPS VI [27,28,29,30].

Type of anaesthesia

General anaesthesia should be undertaken with great care. General anaesthesia is a difficult and potentially high-risk procedure in MPS VI patients, due to the airway management difficulties and cervical cord impingement. Regional anaesthesia has not been reported and is potentially contraindicated [31]. Anaesthesia becomes progressively more difficult with age. Unlike some MPS I and II, some diagnostic procedures like MRI can be performed without general anaesthesia as intellect is preserved and patient cooperation is possible.

Necessary additional diagnostic procedures (preoperative)

Multidisciplinary review is a hallmark of management guidelines for MPS VI [3,6,12,19]. Neurological examination, respiratory function testing, cardiac evaluation and imaging studies are recommended every 12 months (or earlier if symptoms arise) for MPS VI patients [6].

Routine neurological examination with assessment of hyper-reflexia is recommended every 6 months after diagnosis. Cervical stenosis should be evaluated by MRI which is the gold standard to detect compression of the cord, myelopathy and changes of CSF flow [6].

Paediatric cardiology review, including physical examination, electrocardiogram, chest X-ray, and echocardiogram, is necessary. Endurance testing includes the 12 MWT (distance walked in 12 minutes) or 3MSC (3min stair climb) are performed pre and post enzyme replacement therapy and every 12 months. Holter monitoring may be indicated if arrhythmia is suspected [29,30].

Glycosaminoglycans accumulation in the oropharynx and airway combined with the typical dysmorphic features are commonly associated with rhinitis, enlarged tonsil and adenoids, thickening of the epiglottis and narrowing of the trachea and bronchi. Evaluation of pulmonary function by forced spirometry and flow volume expiratory and inspiratory lops should be performed regularly to assess changes in lung volume and obstruction. Comparison to normal values is meaningless but trends are important. Recurrent pneumonia has been reported and preoperative chest X-ray is worthwhile. Resolution of any active respiratory infections prior to surgery is recommended [32].

Obstructive sleep apnoea secondary to upper airway obstruction may lead to failure to thrive, pulmonary hypertension, and behavioural and learning problems. Polysomnography can be used to assess sleep apnoea.

Awake fibre optic naso-endoscopy can be performed to evaluate extent, and severity of airway involvement may aid anaesthetic planning and estimate risk.

Particular preparation for airway management

A survey of MPS patients from the Royal Manchester Children's Hospital demonstrated an overall incidence of difficult intubation of 25% and a failed intubation rate of 8%. Apart from the features that contribute to airway obstruction, patients have craniofacial abnormalities, a short neck, stiffening of the temporomandibular joints, a large tongue, gingival hypertrophy, an anterior larynx and an unstable atlantoaxial joint [33,34].

As difficult bag mask ventilation and difficult or failed intubation is possible; anaesthetic management should be performed by the most experienced anaesthetic team with support of an ENT surgeon. Maintenance of spontaneous respiration is recommended to avoid the 'cannot intubate cannot ventilate' scenario [35].

Neck stabilisation during intubation and during transition from supine to prone position may be required if atlantoaxial instability is present.

Supraglottic airway devices such as laryngeal mask airway have been used successfully and may serve as a conduit for fiberoptic intubation [36]. Video-assisted laryngoscopy has been used successfully in other MPS syndromes but has not been reported in MPS VI. Patients often have thick nasal and oral secretions, hypertrophied turbinates and a narrow nasopharynx making nasal intubation difficult. The use of an oral airway may fail to relieve or even worsen the degree of airway obstruction due to the high elongated position of the epiglottis. Spontaneous breathing induction with a volatile agent, use of a laryngeal mask airway and fibreoptic bronchoscopy to guide intubation have been recommended by Walker et al.

Tracheotomy has been successfully used in two scenarios: 1) to safeguard an anticipated difficult airway prior to a planned surgical procedure, and 2) to treat progressive upper airway obstruction, has been used successfully. An emergency tracheostomy is an extremely difficult procedure in these patients and may not be feasible if the airway cannot be managed. Tracheostomy may not relieve obstruction if there is diffuse tracheal infiltration and tracheal torturosity [37, 38, 39].

Endotracheal extubation should only be undertaken after full reversal of the neuromuscular blockade and if the patient is fully awake, coughing efficiently and breathing adequately. Consider intraoperative steroids (dexamethasone) to help reduce postoperative oral mucosal and tongue swelling. Importantly, extubation should be performed in an area where all the necessary personnel and equipment for re-intubation are available immediately.

Particular preparation for transfusion or administration of blood products

Specific blood products are required for patients' post stem cell transplantation. Leukocyte poor red blood cell products, cytomegalovirus sero-negative, and gamma-irradiated components may be required.

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Particular preparation for anticoagulation

Thrombocytopenia related to galsulfase treatment has been reported but significant bleeding diatheses are rare [40,41]. Dermatan sulfate is structurally related to heparin and has documented antithrombotic properties. Although the excess of dermatan sulfate in MPS VI accumulates primarily in lysosomes and in extracellular matrix (mainly connective tissue); some can be demonstrated to spill into the circulation where it binds to heparin cofactor II. The serum levels of heparin cofactor II-thrombin complex are used as a marker of several of the MPS syndromes. Increased bleeding tendency has been reported by Walker et al.

Particular precautions for positioning, transport or mobilisation

Instability of the atlantoaxial joint and SCC at the upper cervical and thoracolumbar region due to spinal canal narrowing are of prime importance when transporting or moving patients. Awake positioning prior to anaesthesia to find out appropriate position and adequate materials may be of value. Positioning can be difficult due to restricted joint range in elbow, shoulder, hip, knee, and ankles.

Probable interaction between anaesthetic agents and patient's long term medication

There are no reports on interaction between anaesthetic agents and galsulfase.

Anaesthesiologic procedure

Patients with MPS VI should only undergo anaesthesia for imaging or surgery in centres where physicians experienced with the perioperative management of individuals with this disease are available. The parents and patient should receive careful informed anaesthesia consent. A difficult intubation should be assumed and planned for. Review of previous anaesthetic notes is helpful but changes due to disease progression are common. As MPS VI progresses in spite of ERT, techniques that previously had been successful to manage the airway may not be successful for the current procedure. Central neuraxial regional anaesthesia is contraindicated but local or ultrasound guided plexus anaesthesia may reduce analgesic requirements postoperatively [31]. Volatile anaesthetic induction is preferred to maintain spontaneous respiration until the airway is controlled [33].

Providing anaesthesia for MPS VI patients often requires induction in a fully equipped anaesthetic room, with a difficult airway trolley at hand. For imaging procedures induction of anaesthesia in the operating room before transporting the MPS VI patient to the MRI/CT scan suite is advisable. Following major surgery recovery should be performed in the intensive care unit with extubation in a controlled environment.

Particular or additional monitoring

Neurophysiological monitoring with somatosensory evoked potentials (SSEPs) and motor evoked potentials during scoliosis and cervical decompression surgery have been suggested to reduce the risk of spinal cord injury. It is possible to miss motor deficits, and unfortunately parameter changes detected by SSEPs may occur too late to prevent cord damage [42].

Possible complications

- A “cannot intubate - cannot oxygenate” scenario
- Complete airway obstruction, resulting in hypoxemia and cardiac arrest
- Post-obstructive (negative pressure) pulmonary oedema
- Failure to maintain airway after extubation, stridor, upper or lower airway collapse
- Need for urgent reintubation or tracheostomy
- Upper spinal cord injury due to dural thickening, occipito-cervical subarachnoid space narrowing and a dysplastic C1 within foramen magnum.

Postoperative care

There is a risk of upper airway obstruction, and there have been reports of post extubation pulmonary oedema presumably due to forced expiration against a narrowed and thickened glottis.

The degree of postoperative monitoring is dependent on the surgical procedure and preoperative condition of the patient. Intensive care is not mandatory, but intensive care facilities should be on site.

If sleep apnoea is present, use regional local anaesthetic blocks and avoid excessive intraoperative opiates. Continuous oximetry monitoring to detect airway obstruction episodes and desaturation. Consider applying CPAP (Continuous Positive Airway Pressure) or BiPAP (Bilevel Positive Airway Pressure). Postoperative chest physiotherapy has a role in reducing respiratory complications.

Information about emergency-like situations / Differential diagnostics

caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the disease

Acute airway compromise and respiratory failure can be caused by the disease and also as an effect of the anaesthetic.

Loss of lower limb motor evoked potentials (MEPs) during neurosurgery in the prone position may indicate cord ischaemia distal to the operative site. All efforts should be made to increase cord perfusion including increasing blood pressure, surgical repositioning and removal of devices causing cord compression. Failure to regain baseline MEPs should trigger a return to the supine position and awakening of the patient [43,44].

Enzyme replacement infusion reactions include rash, urticaria, headache, hypotension, nausea and vomiting and are often treated with antihistamines, corticosteroids or antipyretics. Confusion may occur with reaction to anaesthetic agents [10].

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Ambulatory anaesthesia

Ambulatory (day case) anaesthesia is not appropriate for MPS VI patients.

Obstetrical anaesthesia

Bacchus et al. reported a pregnant woman with MPS VI who had myelopathy due to compression of the cervical spinal cord by thickened dura. During the last trimester, she had severe neurologic deterioration with spastic quadriparesis and impairment of sphincter function. There was no improvement 2 months after delivery, so a cervical laminectomy and longitudinal splitting of the dura from C-5 to the foramen magnum was done. She experienced good return of function. There are no reports on obstetric analgesia or anaesthesia for patients with MPS VI [45].

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