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Moebius syndrome

orphan anesthesia

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

SUPPLEMENT NR. 14 | 2016

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.

Bisher in A&I publizierte Handlungsempfehlungen finden Sie unter:

www.ai-online.info/Orphsuppl
www.orphananesthesia.eu

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:

www.ai-online.info/Orphsuppl
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orphananesthesia

Anaesthesia recommendations for patients suffering from

Moebius syndrome

Disease name: Moebius syndrome

ICD 10: Q87.0

OMIM: 157900

Synonyms: Congenital facial diplegia (congenital oculofacial paralysis), Möbius syndrome, Moebius sequence, MBS

Moebius syndrome is a rare, nonprogressive neurological disorder (prevalence is estimated to be 0.002% of births) characterized by unilateral or bilateral facial paralysis and defective extraocular eye movements secondary to congenital paresis of the facial (VII) and abducens (VI) cranial nerves. These classic features of the syndrome are often accompanied by hypoglossal (XII), trigeminal (V), glossopharyngeal (IX) and vagal (X) nerve palsies. Affected infants typically present with congenital esotropia and immobile, expressionless facies. Depending on the pattern of cranial nerve involvement, there may be a wide range of clinical expression. Feeding difficulties due to poor coordination of sucking and swallowing may be present with IXth and Xth cranial nerve involvement. This may be associated with dysphagia and retention of oral secretions leading to recurrent bouts of aspiration pneumonia. Inadequate function of the soft palate can also result in dysarthria. Moebius syndrome may also occur in association with various craniofacial (mandibular hypoplasia, microstomia, temporomandibular joint dysfunction, cleft palate, external ear deformities), limb (club foot) and musculoskeletal malformations as well as multiple ophthalmic abnormalities (incomplete eye closure, inability to blink). Other associated manifestations include seizure disorders, congenital heart diseases, hypotonia, hypogonadotropic hypogonadism, hydrosyringomyelia and some degree of mental retardation. It is also associated with prematurity.

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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Disease summary

The cause of Moebius syndrome is unknown, but rhomboencephalic maldevelopment and brainstem ischemia during the first trimester are two possible etiological hypotheses in children with normal karyotype. The list of potential associated teratogenic events has included hyperthermia, trauma, thrombus formation, embolism, haemorrhage, as well as in utero exposure to various medications including misoprostol. Most cases are sporadic, but some familial cases are also known. The inheritance patterns of Moebius sequence are heterogeneous and can be autosomal recessive, autosomal dominant or even X-linked. Some candidate regions and candidate genes (3q21-q22 and 13q12.2-q13) have been described, but no causative gene has yet been confirmed.

The syndrome has most frequently been confused with hereditary congenital facial paresis, which is restricted to involvement of the facial nerve, and no other abnormalities.

Poland-Moebius syndrome is a rare congenital disorder that includes combination features of Poland and Moebius syndromes. Poland syndrome consists of absence of pectoralis major muscle, syndactyly, barchydactyly, and hypoplasia of the hands.

Typical surgery

Anaesthesia for imaging (CAT scan and MRI) and surgical procedures that may include correction of eyes anomalies (e.g. strabismus surgery, ptosis repair, tarsorrhaphy), orthopaedic problems (correction of limb abnormalities), plastic/reconstructive (cleft palate, jaw surgery, facial reanimation surgery), otolaryngological, dental (teeth extractions), or general surgical interventions.

Type of anaesthesia

The potential for problems with aspiration of oral secretions should be remembered, and the use of antisialogogue premedication is recommended.

General anaesthesia is potentially high-risk, due to the airway management difficulties.

Regional anaesthesia per se is not contraindicated, however, positioning problems and scoliosis can affect the success rate negatively.

Necessary additional diagnostic procedures (preoperative)

Although a literature review did not identify reports of an association of cardiomyopathy with Moebius syndrome there are suggestions of myocardial involvement (ventricular septal defect, ductus arteriosus, dextrocardia) thereby emphasizing the consideration of preoperative echocardiography in these patients if one has not been previously obtained.

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Particular preparation for airway management

Craniofacial changes, which can hinder tracheal intubation considerably, are seen in approximately 90% of the patients. Micrognatia, retrognatia, mandibular hypoplasia, and cleft palate are some of the manifestations seen in these patients.

Anaesthesia using a facemask with or without a Guedel airway appeared to be well tolerated and is probably the method of choice where appropriate. Facemask ventilation is usually adequate.

Combination of measures could be used to facilitate intubation, including cricoid pressure, stylettes, gum elastic bougie, two-person technique, changing of the laryngoscope blade, and fiberoptic scope.

In a case of failed intubation the airway could be managed with a laryngeal mask, although its placement could also be difficult (has been reported to have a high failure rate).

Difficult airway equipment should be available.

Particular preparation for transfusion or administration of blood products

None known or reported.

Particular preparation for anticoagulation

None known or reported.

Particular precautions for positioning, transport or mobilisation

Care should be taken to position and protect all affected limbs in the neutral position to reduce the risk of pressure areas or neuropraxia.

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

Although seizures are not a consistent finding in patients with Moebius syndrome, there are reports of associated epilepsy. Anaesthetic care in patients with a seizure disorder should include the recent documentation of serum anticonvulsant concentrations to ensure therapeutic dosing. Continuation of anticonvulsant medications to maintain therapeutic levels intraoperatively is of significant concern as is the immediate reinstitution of chronic seizure therapy during the postoperative period. The induction of hepatic enzymes by certain anticonvulsants may alter the pharmacokinetics and pharmacodynamics of several drugs, including neuromuscular blocking agents (NMBAs). Increasing the intraoperative doses of NMBAs, and certain intravenous anaesthetic induction agents may be necessary with concomitant anticonvulsant therapy.

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Anaesthesiologic procedure

Associated mental retardation and visual and hearing disturbances, both of which may present a major challenge in the communication with and assessment of the patient.

Involvement of the hypoglossal nerve can lead to hypoglossia or ankyloglossia with abnormalities of tongue coordination. These may further increase the likelihood of problems with secretions. The use of antisialogogue premedication is recommended.

Facial paralysis may result in incomplete eye closure and the inability to blink, thereby placing the patient at risk for exposure keratopathy and corneal ulcerations even without anaesthetic care. In such cases, meticulous attention to eye care is suggested.

Induction of anaesthesia can be intravenous or inhalational.

The use of succinylcholine should be avoided because of the potential risk of rhabdomyolysis, hyperkalemia, and malignant hyperthermia. The absolute risk of malignant hyperpyrexia (MH) is unknown. A single case report of fatal MH in an infant with Moebius syndrome exists in the literature to date.

Given the association with abnormal ventilatory control, caution with opioids may be appropriate. Extended respiratory monitoring may be required.

Particular or additional monitoring

Train-of-four monitoring due to hypotonia. Electrodes should be placed at a site, which is not affected by the disease process.

Possible complications

Abnormalities of the orofacial structures are common and as a result may lead to difficulties with intubation.

Potentially increased risk of regurgitation, and aspiration of oral secretions or gastric contents in the perioperative period.

Secretions can cause partial airway obstruction and hypoxaemia.
Respiratory failure secondary to excessive airway secretions could require postoperative mechanical ventilation.

Acute and chronic pulmonary complications, which may result from aspiration.

Abnormalities of ventilatory control (apnoea, hypopnoea) presumably due to associated lesions in the brainstem.

Deformities of upper or lower limbs can be associated with difficulty in securing intravenous access.

Care should be taken to prevent corneal abrasions.

Postoperative care

Paresis of the facial nerve, and the consequent absence of facial expression impair the patient's ability to communicate nonverbally, rendering it difficult to evaluate these patients and assess their pain. Changes in physiological parameters (heart rate and blood pressure) should be used to assess level of analgesia. It is also helpful to enlist the mother's help with an assessment of child's level of comfort.

Hypotonia can also impact on postoperative respiratory function at both the level of the upper airway, and the thoracic musculature and diaphragm. This is especially relevant during the perioperative period when residual anaesthetic agents, and NMBAs may exacerbate poor baseline function leading to respiratory failure.

Continuous postoperative monitoring of respiratory function is recommended.

For postoperative analgesia drugs with limited effects on central respiratory function should be used. Non-opioid analgesics should be used preferentially.

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

Not known or reported.

Ambulatory anaesthesia

The ambulatory surgery should take place at a tertiary care institution with adequate resources, and in selected cases.

Obstetrical anaesthesia

Not reported.

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