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Dyke-Davidoff-Masson syndrome

Ellis-van Creveld syndrome

orphanaesthesia

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

SUPPLEMENT NR. 2 | 2022

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:

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OrphanAnesthesia –

a 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.

Find a survey of the recommendations published until now on:

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orphananesthesia

Anaesthesia recommendations for

Dyke-Davidoff-Masson syndrome

Disease name: Dyke-Davidoff-Masson syndrome

ICD 10: Q04.3

Synonyms: Cerebral hemi-atrophy; Cerebral hemi-hypoplasia

Disease summary: Dyke-Davidoff-Masson syndrome [1] is an extremely rare neurological condition with a greater frequency in the paediatric population [3]. It was described by three clinicians in 1993, Dyke, Davidoff and Masson, who reported [9] patients with facial asymmetry, contralateral hemiparesis, seizures and mental retardation with pneumatoencephalographic findings on skull X-ray. There is a slight male [7] preponderance. Adult cases [10] have also been reported, though very infrequently. The main causes of this rare syndrome are either congenital (in utero vascular occlusion) or acquired (perinatal hypoxia, intra-cranial haemorrhage and infections). Brain imaging [2] in the form of CT and MRI scans are required for diagnosis.

Literature regarding anaesthetic considerations for this syndrome is sparse.

Differential diagnoses [9] include Sturge-Weber syndrome, Fishman syndrome, Basal cell germinoma, Silver-Russell syndrome, Linear nevus syndrome and Rasmussen encephalitis.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

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Typical surgery

Patients with Dyke-Davidoff-Masson syndrome can present for any type of elective or emergency surgery to the attending anaesthesiologist. Generally, these are paediatric patients [4]. Because of the occurrence of recurrent seizures and hemiparesis, they are prone to falls and hence develop orthopaedic injuries. Surgery may be required for fractures, non-unions, contractures or, sometimes, neurosurgical operations. Rarely, a few patients can undergo hemispherectomy for treatment-resistant seizures (success rate of 85 % in carefully selected patients).

Type of anaesthesia

Anaesthesia for patients with Dyke-Davidoff-Masson syndrome can be quite challenging. There is no literature regarding anaesthetic considerations for this rare syndrome. In the presence of mental retardation, patient cooperation for regional anaesthesia and intravenous cannulation can be quite difficult to obtain. Both general and regional anaesthesia can be given with proper precautions. The preferred technique in such patients would be general anaesthesia, followed by ultrasound-guided regional nerve block.

In patients with developmental delays, venous cannulation can be challenging. The use of ultrasound-guidance for vascular access is recommended to improve success rates. Spinal deformities can also compound central neuraxial block performance and effect. Treatment medications taken for convulsions, hemiparesis and learning difficulties can have interactions with anaesthetic agents. Most anti-convulsants are enzyme inducers. Some patients may be taking drugs for psychiatric disorders like schizophrenia [8].

Necessary additional pre-operative testing (beside standard care)

If not already done before, neuroimaging [5] is a must for all these cases, which will show atrophy or hypoplasia of one of the cerebral hemispheres. History about brain insult in the foetal or early childhood period should be elicited, as well as maternal thrombophilia (in case of congenital cases) Other causes of mental retardation must be ruled out. An objective examination for the cause of seizures must be done and adequate epilepsy management must be started pre-operatively. Besides standard pre-anaesthetic investigations, skull radiograph, contrast-computed tomographic brain scan (CECT) and electro-encephalography may be considered for evaluation. MRI (magnetic resonance imaging) of the brain can highlight changes in the cerebral hemisphere as well as bony structures, thus differentiating between acquired and congenital Dyke-Davidoff-Masson syndromes: the heterolateral brain structures are shifted towards the affected side in case of a congenital cause. Complete neurological assessment must be done to ascertain the level of hemiparesis (grade of motor/sensory loss). Serum anticonvulsant levels may be done in selected cases to ascertain adequate anti-epileptic medication level as well as to rule out their toxicity. Valproate can interfere with coagulation (fibrinogen) and a coagulation screening should ideally be performed before surgery at risk of bleeding. Special attention must be given to renal function tests and serum electrolytes.

This is part of the diagnostic setup of the syndrome: We suggest adding a section on diagnosis to include all these points. This should not be repeated before anaesthesia if the patient's medical file is ok.

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Particular preparation for airway management

There is a possibility of difficult airway in view of facial asymmetry, ipsilateral hypertrophy of skull and sinus enlargement. Due to learning difficulties and mental retardation, awake intubation is not possible due to lack of patient cooperation. Inhalational anaesthesia or Intravenous sedation with preservation of spontaneous ventilation should be used for endotracheal intubation if a difficult airway is suspected or foreseen. Mask ventilation may be difficult due to improper or inadequate mask seal. Aspiration prophylaxis may be given in patients with recurrent or intractable seizures [6] and mental retardation. A difficult airway cart must be kept ready at all times and standard ASA guidelines must be followed for airway management.

Particular preparation for transfusion or administration of blood products

There are no known particular recommendations regarding transfusion or administration of blood products.

Particular preparation for anticoagulation

There is no known specific preparation for anticoagulation. Caution needs to be exercised in patients taking anticonvulsants and warfarin in view of enzyme induction.

Particular precautions for positioning, transportation and mobilisation

Peri-operative positioning needs special attention as these patients are mentally challenged and may have several facial/skull abnormalities. Documentation of pre-existing neuropathies must be done. All pressure points must be padded. Transportation and mobilisation must be gentle to prevent musculo-skeletal injuries.

Interactions of chronic disease and anaesthesia medications

There are no known disease-specific interactions with anaesthetic drugs. However, the medications taken by these patients can have interactions with anaesthetic agents, necessitating dose adjustments.

Anaesthetic procedure

Standard general anaesthesia followed by administration of regional nerve blocks (USG-guided) when appropriate is beneficial in these patients. Aspiration prophylaxis, preparation for difficult airway, securing a good venous access, ensuring adequate oxygenation and ventilation are the cornerstones of successful outcome. These children can have exaggerated separation anxiety and may have agitated behaviour [11] pre-operatively, leading to difficulty in induction. Extubation must also be cautious and expedited only after ensuring wakefulness and adequate tidal spontaneous ventilation. Post-operative care must

be in a high-dependency care unit with requisite monitoring and involvement of the care-givers.

Particular or additional monitoring

Apart from standard ASA monitors, peripheral neuromuscular monitoring and depth of anaesthesia monitor are recommended to prevent delayed reversal or awakening from anaesthesia. Due to the underlying cerebral anomalies and increased risk of convulsions, adequate depth of anaesthesia monitoring includes measuring its awake values (which can be challenging in this population) and interpreting any brisk increase as a possible sign of seizures. Invasive haemodynamic monitoring may be indicated in patients with other associated cardio-pulmonary abnormalities. Temperature monitoring must be meticulous to prevent the development of hypothermia in these vulnerable patients.

Possible complications

Complications can be disease-, anaesthesia- or procedure-related or due to surgical reasons. Special attention must be given to prevent hypoxia or hypercarbia, as these can trigger seizures. Some patients may develop emergence delirium or agitation and require prolonged hospital stay. The incidence of PONV (post-operative nausea vomiting) can be theoretically high, although no randomised studies have been done in this population. Pharmacologic PONV prophylaxis must be given in all patients. These patients must be dealt with compassion in view of their learning difficulties.

Post-operative care

These patients must be monitored in a high-dependency unit in the post-operative period. Vigilant observation must be done for seizures and anticonvulsant medications must be continued. Supplemental oxygen, adequate analgesia, psychological support and prevention of hypothermia must be ensured.

Disease-related acute problems and effect on anaesthesia and recovery

There are no specific interactions between disease-related acute problems and effect on anaesthesia and recovery. There could be delayed recovery from general anaesthesia due to the prolonged effects of anaesthetic agents, electrolytic disturbances or a post-critical state following seizures under anaesthesia. It is necessary to use neuromuscular monitoring to ensure adequate reversal from muscle relaxants.

Ambulatory anaesthesia

Ambulatory anaesthesia is only advisable for short non-painful procedures. Standard precautions of postoperative care and peri-operative monitoring is advised. The presence of an expert care giver must be ensured before discharge. It is preferable to avoid day-care surgeries in view of seizure disorders, learning difficulties and maxillo-facial deformities.

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Obstetrical anaesthesia

Literature is sparse regarding obstetric anaesthesia in patients with DDMS. Aspiration prophylaxis, adequate oxygenation, avoidance of supine-hypotension syndrome, continuation of anticonvulsants, and difficult airway preparedness must be achieved. It must be remembered that administering regional anaesthesia (central neuraxial block) may be extremely difficult in these patients due to lack of co-operation. Proper antenatal counselling must be done to develop a rapport with the patient to seek her co-operation for regional block, as it is much safer than general anaesthesia in an obstetric patient.

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Please note that this recommendation has not been reviewed by an anaesthesiologist and a disease expert but by two anaesthesiologists instead.

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