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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.

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:

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orphananesthesia

Anaesthesia recommendations for patients suffering from

McCune-Albright syndrome

Disease name: McCune-Albright syndrome

ICD 10: Q78.1

Synonyms: Polyostotic fibrous dysplasia, MAS, Albright syndrome, osteitis fibrosa disseminata, PFD, precocious puberty with polyostotic fibrosis and abnormal pigmentation, POFD

McCune-Albright syndrome (MAS) is a disease that affects the skin, bones and endocrine tissues. The incidence of MAS is rare, affecting 1 in 100,000 to 1 in 1,000,000 people globally. It is not inherited, but results from a random mutation in the *GNAS* gene, an over-activation of adenylate cyclase and hormonal dysregulation. It classically presents with the presence of at least two of its three most common findings; polyostotic fibrous dysplasia (abnormally fragile bones that easily fracture), hyperpigmented skin patches (café au lait spots), and endocrine dysfunction. Acromegaly, pathologic fractures, spinal instability, scoliosis, hyperthyroidism, hypophosphatemia, liver disease, obstructive sleep apnea, neonatal Cushing's syndrome, neuropathy, hypertension and arrhythmia can also accompany MAS. Airway management is complicated by distortion of the face and airway. Abnormal bony growth can cause skeletal fractures and spinal instability, which can increase the risk of injury related to patient positioning. Endocrinopathies may also contribute to perioperative cardiac arrhythmias and haemodynamic instability.

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

▶ **Citation:** Moran K, Straka B, Rankin D: McCune-Albright syndrome. Anästh Intensivmed 2017;58:S515-522. DOI: 10.19224/ai2017.S515

Typical surgery

Surgeries common to MAS patients include needle biopsy, transsphenoidal hypophysectomy, internal fixation following bone grafting, scoliosis correction, thyroidectomy and adrenalectomy. Bone grafting has been shown to be ineffective in treating fibrous dysplasia in young patients, and may become less common, particularly in children.

Type of anaesthesia

General and regional anaesthesia can be safely administered. However, the severity of the disease and its effect on various organ systems may affect the anaesthetic plan. An increased risk for difficult mask ventilation and intubation due to acromegaly and bony malformation should be considered. During regional or local anaesthesia, airway concerns may limit the level of sedation that can be safely delivered. In addition, the risks of positioning injury due to abnormal bone formation may be reduced when general anaesthesia can be avoided. For example, the presence of facial deformities requires particular attention to the face and eyes during surgery under general anaesthesia in the prone position while an awake patient under regional anaesthesia is able to adjust their position if compression were to occur.

Important anaesthetic considerations include determining which of the endocrine systems have been affected and evaluation of the extent of hormonal derangement. Avoidance of general anaesthesia may be helpful when severe derangements of thyroid hormone, cortisol or electrolytes are present.

Necessary additional diagnostic procedures (preoperative)

When MAS patients present with Cushing's syndrome, it is important to screen for poorly controlled hypertension, elevated glucose levels, and hypokalemia. A baseline serum chemistry study should be used to evaluate for sodium, calcium, phosphorus, potassium, and glucose abnormalities. Cushing syndrome in MAS patients is typically only seen in neonates or small children since most occurrences are either treated with adrenalectomy or managed medically until spontaneous resolution occurs in early childhood. Verification that MAS patients are under the care of an endocrinologist who has followed appropriate screening guidelines will help reduce the likelihood of an undiagnosed endocrinopathy.

Thyroid function tests should be evaluated if there are clinical concerns for thyroid dysfunction. Most often hyperthyroidism is the primary concern. Ideally, patients should be adequately treated to ensure that a euthyroid state is achieved prior to elective surgery. If hyperthyroidism is left uncorrected, thyroid storm may occur. In addition, laboratory evaluation can assist in the preoperative recognition of hypophosphatemia. Additional screening tests for acromegaly and hyperprolactinemia may also be helpful.

Patients with MAS can have accompanying cardiac disease including atrial fibrillation, arrhythmias and non-ischemic cardiomyopathy, particularly in cases involving acromegaly. Untreated hyperthyroidism and hypertension, in particular, can lead to cardiac dysfunction. Preoperative electrocardiogram (ECG) should be obtained as a baseline assessment and to rule out arrhythmia or other pathologies, particularly when electrolyte abnormalities are present. In addition to a screening ECG, patients that demonstrate poor exercise capacity in the absence of ischemic heart disease may require echocardiography.

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Screening for obstructive sleep apnoea (OSA) should always be considered in patients with acromegaly since soft-tissue enlargement of the tongue and laryngeal structures can lead to obstruction. Also, OSA risk factors may influence pain management strategies.

Some patients with MAS may present with severe scoliosis. In extreme cases, preoperative pulmonary function tests may identify restrictive lung disease resulting from the spinal abnormality. Since restrictive pulmonary disease increases the risk of postoperative pulmonary dysfunction, identification of restrictive lung disease may guide perioperative fluid management, influence ventilation strategies, and potentially avoid postoperative pulmonary complications.

Particular preparation for airway management

Patients with McCune-Albright syndrome have a variety of physical deformities that can complicate airway management. Acromegaly, which commonly results from MAS associated endocrine hyperfunction, is associated with craniofacial abnormalities such as macroglossia and macrognathia. These changes can distort airway anatomy and complicate mask ventilation and intubation. Since acromegaly can result in soft tissue hypertrophy, including thickening and widening of the larynx, consider consultation with an otolaryngologist to determine if an advanced airway examination is required.

To further complicate airway management, polyostotic fibrous dysplasia results in bony changes that affect oral opening and can put the patient at risk for mandibular and maxillary fracture. Soft tissue hypertrophy can also affect visualization. Obesity, bony hypertrophy and the classic "buffalo hump" associated with Cushing's syndrome can distort optimal airway positioning. In many cases, flexible fiber optic laryngoscopy may be required for safe airway management. It is important to note that pressure on the maxilla or mandible during direct or fiberoptic laryngoscopy can result in a pathologic fracture in areas affected by bony dysplasia.

Precaution must also be taken when manipulating the head and neck of MAS patients. Skull base fibrous dysplasia and spinal instability from a large head and spinal compression fractures have been described.

Particular preparation for transfusion or administration of blood products

Standard transfusion practices should be applied to patients with MAS.

Particular preparation for anticoagulation

Standard anticoagulation practices apply to patients with MAS. Of note, care should be exercised when placing compression hosiery or automated sequential compression devices for deep vein thrombosis prophylaxis.

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Particular precautions for positioning, transport or mobilization

Increased bone fragility requires special attention to ensure adequate padding and positioning. Bony growth that presses on peripheral nerves can increase the likelihood of neuropathic injury.

Patients with acromegaly often have an enlarged and irregularly shaped head. If prone positioning is required, head positioning devices may require extra padding or alteration to avoid pressure injury. The enlarged head may also impede neutral alignment of the cervical spine. Particular attention should be paid to the eyes and any facial deformities in order to avoid compression due to inadequate padding in the prone position.

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

Patients who have received a bilateral adrenalectomy, or those with a history of resolved neonatal Cushings, may experience adrenal insufficiency during the perioperative period. Stress dose steroids should be considered for these patients. Octreotide combined with steroids administered perioperatively can affect blood glucose levels, which should be followed closely.

Anaesthesiologic procedure

Induction of anaesthesia will often require advanced airway techniques such as awake fiberoptic intubation or backup airway devices.

During induction, close monitoring for arrhythmia due to electrolyte imbalance and endocrinopathy is warranted. Sudden cardiac death has also been reported in these patients.

Most anaesthetic agents including volatile agents, propofol, depolarizing and non-depolarizing muscle relaxants, local anaesthetics and opioids have been used without complications. In patients where cortisol dysfunction is suspected, avoidance of etomidate may be warranted because of its propensity to cause adrenal suppression.

Particular or additional monitoring

Patients should be monitored according to the risk of haemodynamic instability that accompanies the surgery and their baseline cardiac function, including their risk for arrhythmia. Arterial or central venous access should be considered if the patient has experienced arrhythmias, cardiomyopathy or if heavy blood loss or significant fluid shifts are expected.

Since MAS is associated with Cushing's syndrome, electrolyte abnormalities, and hyperthyroidism, these patients should be considered high risk for arrhythmia and haemodynamic instability.

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Possible complications

Venous access can be difficult in patients with vascular fragility due to Cushing's syndrome.

Preparation for airway emergencies is essential in patients with acromegaly. Distorted anatomy, cervical instability, and fragile bone structure must be considered at both intubation and extubation. Consider developing a plan for re-securing a difficult airway in the setting of failed extubation.

Increased bone fragility places MAS patients at risk for mandible and face fractures during airway manipulation. Meticulous positioning is also important to avoid skeletal fractures that can result from bony fibrous dysplasia. These patients are also at risk for neuropathy from bony compression.

Patients with McCune-Albright syndrome are at risk for haemodynamic instability from thyroid storm, arrhythmia, electrolyte abnormality or steroid imbalance. These risks are increased when urgent or emergent surgery does not permit complete evaluation and optimization of endocrinopathies and cardiac function. Electrolyte imbalance may also predispose patients to arrhythmias.

In patients with Cushing's syndrome, the potential for inadequate adrenal response to stress must be considered. These patients may require perioperative steroid administration.

Postoperative care

While recovery in an intensive care setting is not required for all patients with McCune-Albright syndrome, it should be considered for patients with significant endocrine dysfunction, electrolyte imbalance, cardiomyopathy, airway management issues, or whose surgical course merits close postoperative monitoring.

Thyroid storm, is of particular concern and can occur 6-18 hours after surgery.

Peripheral nerve injuries from inappropriate positioning can also occur during the recovery period. Neutral positioning, awareness of bony outgrowths at pressure points and limb positioning on padded areas should be maintained.

Consider postoperative monitoring with telemetry and pulse oximetry since patients with MAS are at increased risk for obstructive sleep pathology, especially when sedated.

Information about emergency-like situations / Differential diagnostics

Hyperthermia associated with intraoperative thyroid storm has been misinterpreted as malignant hyperthermia (MH). Interestingly, it has been suggested that dantrolene may also successfully treat thyroid related hyperthermia.

Airway difficulty must always be considered in patients with craniofacial abnormalities such as those found in McCune-Albright syndrome. Backup airway plans and surgical presence are recommended in cases where the potential for an invasive airway exists. Preoperative evaluation by an otolaryngologist may demonstrate soft tissue hypertrophy and thickening or widening of the larynx. Extubating a patient with a proven difficult intubation may require additional planning and preparation for possible reintubation.

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Ambulatory anaesthesia

Surgeries on MAS patients in the ambulatory setting should be limited to those without significant endocrine system impairment, cardiac dysfunction, electrolyte imbalance or airway concerns who are receiving low risk surgery. Concern for postoperative obstructive sleep apnea may also preclude surgery in the ambulatory setting.

Obstetrical anaesthesia

While precocious puberty is considered a hallmark of MAS, women with this disease have demonstrated the ability to become pregnant. In addition to airway changes associated with acromegaly and fibrous dysplasia, physiologic changes of pregnancy will further distort airway anatomy. Careful evaluation and planning is necessary. Altered morphology from scoliosis, acromegaly and Cushing's syndrome may also complicate the placement of neuraxial anaesthesia.

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Literature and internet links

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