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Organ: Deutsche Interdisziplinäre Vereinigung für Intensiv- und Notfallmedizin e.V. (DIVI)

Rett syndrome

ROHHAD syndrome

orphananesthesia

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

SUPPLEMENT NR. 6 | 2018

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
www.orphananesthesia.eu

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orphananesthesia

Anaesthesia recommendations for patients suffering from

ROHHAD syndrome

Disease name: ROHHAD syndrome

ICD 10: -

Synonyms: Rapid onset obesity, hypoventilation, hypothalamic dysfunction, and autonomic dysfunction

ROHHAD is a clinical entity with a median age of 3 years at onset characterised by sudden onset of dramatic weight gain, dysautonomia, and pulmonary complications. These include alveolar hypoventilation, obstructive sleep apnoea, and decreased central responsiveness to carbon dioxide concentrations. There are also several endocrinopathies associated with this disorder including hypernatraemia, hyperprolactinemia, hypothyroidism, and diabetes insipidus. Additional features of the disease include behavioural and mood disorders, as well as seizures and learning impairment. There is an associated entity known as ROHHADNET with a predisposition towards the development of neuroectodermal tumours. Therefore, all patients with known ROHHAD syndrome should be carefully screened for the presence of these tumours.

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

▶ **Citation:** Chandrakantan A: ROHHAD syndrome. Anästh Intensivmed 2018;59:S125-S130.
DOI: 10.19224/ai2018.S125

Typical surgery

Depends on presentation of disease. May vary from GI procedures from Hirschprung's disease, surgeries for abnormal ventilation (diaphragmatic pacing, tracheostomy placement), and dysautonomia (permanent pacemaker placement). May also be present for any surgery in the typical array of paediatric surgeries.

Type of anaesthesia

There is no contraindication to general or regional anaesthesia, although patients must be monitored carefully. In particular, in patients with autonomic dysfunction, a potential sympathetic blockade resulting from regional anaesthesia requires careful control of blood pressure. Consequentially, a neuraxial blockade should be cautiously administered to these patients.

Necessary additional diagnostic procedures (preoperative)

If known, evaluation for all associated disorders. This includes an assessment of gastric emptying (gastroparesis), cardiac abnormalities, including the presence or absence of a pacemaker. Other concerns include electrolyte abnormalities, seizure disorders, untreated endocrinopathies, behavioural disorders, and possible thermal dysregulation. At the very minimum, the anaesthesiologist should perform a detailed history and physical examination to ensure adequate treatment of existing disorders, and identify any other conditions which need optimisation prior to surgery.

In addition, children should be evaluated for associated disorders that have not been diagnosed. Especially the endocrinopathies, the stress of surgery may uncover unknown and untreated disorders. These include:

- Abnormal hypothalamic-pituitary-adrenal axis
- Central hypothyroidism
- Impaired glucose tolerance or diabetes mellitus
- Diabetes insipidus

In all patients with ROHHAD preoperative pulmonary evaluation is strongly recommended (i.e., an evaluation of sleep-disordered breathing and the measurements of respiratory function) in order to optimise the patient's respiratory status before surgery. When respiratory function measurements and/or sleep studies are abnormal, non-invasive ventilation (NIV) may be indicated. Consequently, these patients should be trained in NIV before surgery and assisted with NIV during sedation, regional anaesthesia and in the postoperative period.

Particular preparation for airway management

An increased incidence of difficult airway has not been identified in these patients. There is a strong association with postoperative apnoea, ventilatory and anaesthetic management should therefore be identified early and used judiciously. However, as suggested by multiple

authors, morbid obesity and the high incidence of OSA makes these patients high risk for difficult mask ventilation.

In patients who possess tracheostomies, there are situations in which switching to a cuffed endotracheal tube may be optimal.

Particular preparation for transfusion or administration of blood products

No unique concerns applying to this patient population are known.

Particular preparation for anticoagulation

Despite the prevalence of endocrinopathies in these children, there is no contraindication to the use of anticoagulation in these patients.

Particular precautions for positioning, transport or mobilisation

These children can be morbidly obese and may have devices including tracheostomy tubes, PEG tubes for feeding etc. Therefore caution is warranted when mobilising these patients.

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

Many of these patients are on seizure medications, and other psychotropic or mood enhancing medications. Therefore, the anaesthesiologist is advised to study the medication list, ask when the patient last took the medication, and to plan the anaesthetic accordingly.

Anaesthesiologic procedure

As noted by Chandrakantan and Poulton, the judicious use of premedication with minimal respiratory effects, such as benzodiapenes, in children with behavioural disorders may be of benefit. These have been used safely.

Intraoperatively, the use of inhalational agents has been safe and documented in these children. Additionally, the use of intravenous agents with short half-lives and minimal respiratory effects is advisable. Agents that have been used safely intraoperatively include ketamine and dexmedetomidine. Non-depolarising muscle relaxants have been used safely. There is no known contraindication to the use of succinylcholine.

Particular or additional monitoring

Postoperatively, these children are prone towards prolonged apnoea and carbon dioxide retention. Therefore, careful postoperative care includes respiratory monitoring with end-tidal

CO_2 monitoring, blood pressure monitoring for any lability due to dysautonomia, and the judicious use of pain medications to control pain, but not to cause respiratory depression.

It has been suggested that opioids be avoided in order to avoid respiratory depression and that non-opioid pain adjuvants should be used in their entirety to control pain. While this seems correct intuitively, there is not enough data to support this view.

Possible complications

Most of the complications usually occur when the disorder is not thought of or entertained until the child has prolonged postoperative apnoea. Blood pressure lability from dysautonomia is also a possible complication. Careful attention to thermal dysregulation should also be monitored by use of temperature-measurement devices.

Postoperative care

As noted above. Patients who were on non-invasive ventilation prior to surgery should continue their ventilation postoperatively. PICU admission should be strongly considered to closely monitor postoperative respiratory parameters.

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 diseases, e.g.:

Prolonged postoperative apnoea needs to be distinguished from prolonged anaesthetic medication effect, which is very difficult to do in practice. Hence the suggestion of the use of medications with minimal respiratory depressant effects and short half-lives to minimise the probability of the latter.

Ambulatory anaesthesia

Although there are no data to suggest that ambulatory anaesthesia is intrinsically dangerous in these patients, the nature of the disease does suggest that the patients should be hospitalised for at least 24 hours postoperatively to minimise any lingering medication related issues.

Obstetrical anaesthesia

No data currently exist on this subject.

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Literature and internet-links

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