orphanaesthesia

Anaesthesia recommendations for

ROHHAD

Disease name: ROHHAD

ICD 10: --

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

Disease summary: ROHHAD is a clinical entity with a median age of 3 years at onset characterized by sudden onset of dramatic weight gain, dysautonomia, and pulmonary complications. These include alveolar hypoventilation, obstructive sleep apnea, and decreased central responsiveness to carbon dioxide concentrations. There are also several endocrinopathies associated with this disorder including hypernatremia, 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 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

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 pre-operative testing (beside standard care)

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 optimization 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 optimize 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

There has not been an increased identified incidence of difficult airway in these patients. There is a strong association with postoperative apnoea, therefore ventilatory and anaesthetic management should 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 exists situations in which switching to a cuffed endotracheal tube may be optimal.

Particular preparation for transfusion or administration of blood products

No unique concerns to this patient population 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, transportation and mobilisation

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

Interactions of chronic disease and anaesthesia medications

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 their anaesthetic accordingly.

Anaesthetic 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 dexmetomidine. Non depolarizing 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 CO2 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 adjuvant should be used in their entirety to control pain. While this seems intuitively correct, 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.

Post-operative 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.

Disease-related acute problems and effect on anaesthesia and recovery

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

Ambulatory anaesthesia

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

Obstetrical anaesthesia

No data exists currently on this subject.

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