orphanaesthesia

Anesthesia recommendations for patients suffering from

Prader-Willi syndrome

Disease name: Prader-Willi syndrome

ICD 10: Q87.1

Synonyms: Prader-Labhardt-Willi syndrome

Prader-Willi syndrome is a rare genetic disorder characterized by hypothalamic-pituitary abnormalities with severe hypotonia during the neonatal period and during the first two years of life, hyperphagia with a risk of morbid obesity during infancy and adulthood, learning difficulties and behavioral problems or severe psychiatric problems. The disease affects 1/25,000 births. The severe hypotonia at birth, which leads to sucking and swallowing problems and delayed psychomotor development, partially improves with age. Characteristic facial features (a narrow forehead, almond-shaped eyes, a thin upper lip and down- turned mouth), as well as very small hands and feet, are frequently observed. After this initial phase, the most striking signs appear including hyperphagia and absence of satiety often leading to severe obesity in affected children as young as two years of age. The situation may deteriorate quickly without adequate outside controls and obesity is a major factor influencing morbidity and mortality in these patients. Other associated endocrine abnormalities contribute to the clinical picture of short stature due to a growth hormone (GH) deficiency and incomplete pubertal development. Regularly, a decreased bone mineral density can be found without changed metabolism of calcium, phosphate, Vitamin D or parathyroid hormone. The degree of cognitive dysfunction varies widely from child to child. It is associated with learning disabilities, and impaired speech and language development that are aggravated further by psychological and behavioral troubles.

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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

The disease is clinically and genetically heterogeneous. It is caused by anomalies involving the critical region of chromosome 15 (15q11-q13). The expert consensus is that diagnosis should be based on clinical criteria (Holm's criteria of 1993, revised in 2001) with confirmation by genetic analysis. Most cases are sporadic and familial recurrence is rare information that should be provided by genetic counseling. Management should be global and multidisciplinary. Early diagnosis, early multidisciplinary care and GH treatment have greatly improved the quality of life of affected children. There are currently no long-term data on the effect of GH treatment in adults, particularly concerning its effect on the behavioral problems and degree of autonomy obtained. In adults, complications linked to obesity and the issue of autonomy continue to pose important problems

Typical surgery

Orthopedic surgery (e.g. spinal surgery for scoliosis); squint correction; cleft lip and palatal repair; dental treatment due to tooth decay; Orchidopexy (cryptorchism).

Type of anaesthesia

Both general and regional anesthesia are challenging: using general anesthesia may result in difficult airway management, landmarks for regional anesthesia may be obscured due to morbid obesity.

Anaesthetic concerns of PWS include morbid obesity and sleep apnea, difficult intravenous access, the potential for difficulties with airway management, risk for perioperative respiratory failure, primary myocardial involvement, aggressive and at times violent behavior, convulsions, and disturbances in thermoregulation as well as glucose intolerance.

Necessary additional diagnostic procedures (preoperative)

Appropriate preoperative evaluation may depend on the presence of co-morbid conditions include in obesity-related complications - cardiovascular problems, diabetes mellitus, hypertension, sleep apnea (**NOTE**: predisposition to cardiac- vascular complications independent of obesity).

- state of nutrition and hydration
- body mass index, neck circumference
- dental issues
- salivary composition

Take care of a thorough evaluation of the patient´s past and current respiratory status including Polysomnography as a part of an individualized preoperative workup in order to establish a baseline and identify those with severe OSA (as well as postoperative admission to the ICU for monitoring of respiratory status). The recordings include thoracic and abdominal breathing movements, nasal airflow, TC-PO_2, TC-CO_2, oxygen saturation, EEG, EOG (Electrooculography) and ECG.

Children with PWS frequently suffer from restrictive lung disease because of hypotonia, obesity and kyphoscoliosis.

Preoperative evaluation with 12 lead ECG and echocardiography may be indicated in selected patients).

Particular preparation for airway management

In the perioperative management of a patient with Prader- Willi syndrome, special attention must be paid to the abnormalities in the upper and lower respiratory systems. Airway management may be complicated by the frequent association of poor dentition, micrognathia, palatal abnormalities, and limited neck mobility. The appropriate equipment to deal with the ”cannot intubate/cannot ventilate” scenario should be available. Also in patients in whom regional anesthesia will be performed, there should be ready access to such equipment.

Intraoperative and postoperative respiratory compromise has also been a frequent perioperative problem in patients with PWS. These may include stridor, oxygen desaturation, hypercapnia, and intermittent bronchospasm with the need to use high peak inspiratory pressure.

Particular preparation for transfusion or administration of blood products

Not reported.

Particular preparation for anticoagulation

Not reported.

Particular precautions for positioning, transport or mobilisation

Features that may have an impact on perioperative care include mental retardation with the propensity for aggressive behaviour.

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

There no particular interactions, because there is no specific medication. Interactions depend on the presence of comorbid conditions and its drug therapy.

Anaesthesiologic procedure

Patients with PWS are presenting with muscular hypotension and therefore predisposed to gastric aspiration. The decreased motility of the gastrointestinal tract further heightens the risk of perioperative aspiration, and the sequel of aspiration may be more severe due to limited pulmonary reserves. The physiological set point of vomiting in PWS is abnormal, and these patients have a reduced tendency to vomit. Certainly the obese body habitus may be

associated with a higher than normal incidence of hiatal hernia and increased intra-abdominal pressure. The perioperative aspiration risk is further exacerbated by a high incidence of rumination and lowered esophageal sphincter tone. Rumination may contribute to the development of dental caries. Efforts should be made to reduce gastric acid secretion, increase intestinal motility, neutralize stomach contents, use body position to utilize gravity to reduce the tendency for passive regurgitation, secure the airway rapidly, decompress the stomach, and extubated the trachea cautiously. By these means, surgical morbidity and mortality related to aspiration of gastric contents may be reduced.

- nil per os for 6 hours
- peripheral intravenous cannula, topical anesthetic cream should be used before
- ranitidine and metoclopramide intravenously, 1hour prior to anesthesia
- careful premedication with a sedative
- routine monitoring
- rapid sequence induction

Regional anesthetic techniques play a role in such patients by eliminating the need for general anesthesia and its perioperative risks. Regional anesthesia can be helpful to provide intraoperative anesthetic care and postoperative analgesia as a means of limiting the need for opioids and general anesthetic agents. Both neuraxial techniques (epidural or spinal anesthesia) and peripheral nerve blockade may be considered in these patients. Nevertheless, landmarks for regional anesthesia may be obscured due to morbid obesity and the use of ultrasound is recommended to facilitate placement of the block and limit the incidence of complications.

A risk benefit ratio has to be considered when using regional anesthesia plus sedation in patients with PWS who may be at higher risk of aspiration than the general pediatric population. Sedation will likely be required not only for block placement, but also to ensure a cooperative patient during the surgical procedure.

Moreover, the syndrome may result in a prolonged and exaggerated response to every sedative and analgesic agent. It has been suggested that regional techniques and NSAIDs beneficial as a means of limiting opioid use in the immediate postoperative period.

The use of non-depolarizing neuromuscular blocking agents for muscle relaxation should be restricted because of potential for long- lasting neuromuscular blockade. Hypotonia has prompted some authors to caution against the use of neuromuscular blocking agents. However, several reports have demonstrated the safe use of various non-depolarizing neuromuscular blocking agents including pancuronium, atracurium, vecuronium, and rocuronium without evidence for prolonged effects. Neuromuscular monitoring is necessary and residual neuromuscular blockade should be promptly antagonized with neostigmine and glycopyrrolate. Although they would suggest caution with the use of succinylcholine in the presence of hypotonia given the theoretical risk of an exaggerated hyperkalemic response, several of the reports have also demonstrated the safe use of succinylcholine in patients with PWS.

Some authors chose ketamine for its limited effects on respiratory function as well as its ability to provide both sedation and analgesia. Especially in older children, ketamine should be coadministered with either propofol or a benzodiazepine to limit the potential of emergence phenomena. Ketamine can be used with caution as its effects on the seizure threshold are controversial.

Propofol, isoflurane, sevoflurane and N2O have been used for the maintenance of anaesthesia.

Other common problems include difficult venous puncture, food-seeking behavior, disturbances in thermoregulation, diabetes mellitus, arrhythmia, and cor pulmonale.

Particular or additional monitoring

Monitoring of the neuromuscular blockade is recommended.

Blood glucose and body temperature should be monitored carefully.

Additional monitoring depends on identification of elevated risk for intraoperative or postoperative problems due to comorbidity.

Possible complications

PWS patients tend to develop severe respiratory infections as a result of aspiration due to hypotonia leading to poor pharyngeal coordination, chronic aspiration and a weak cough.

Strategy for the prevention of perioperative complications for PWS patients:

- elective surgery should be postponed until complete recovery from a URI
- special attention must be paid to those patients complicated with severe OSA
- the increased risk of aspiration and postoperative apnea should be considered.

Postoperative care

For close observation and therapy postoperative intensive care medicine is highly recommended to prevent sleep-related respiratory complications – even if neither narcotics nor intermediate or long lasting neuromuscular blocking agents are administered.

Postoperatively, emergence often is slow and accompanied by upper airway obstruction and snoring. Complications may include an increased number of apneic episodes, higher hypercapnic thresholds, obstructive episodes, brochospasm and oxygen requirement.

Wound healing and mobilization can be slow.

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.:

The accompanying morbid obesity, OSA, pathologic central control of ventilation and excessive daytime sleepiness seems to be characteristic for PWS, and may be related to problems with the sleep-wake rhythm and hypothalamic dysfunction.

Patients with PWS present with Mental retardation with the propensity for aggressive behavior, psychological disturbances, and obsessive behavioral problems.

In later childhood and adolescence, neurologic manifestations, endocrine signs and dysmetabolic abnormalities can occur.

Additional psychological disturbances, mental retardation and obsessive behavioral problems are frequent findings.

Cardiac conduction defects and primary myocardial involvement have been noted and convulsions are common.

Ambulatory anaesthesia

Ambulatory anesthesia is not recommended.

Obstetrical anaesthesia

Not reported.

Literature and internet-links

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