orphanaesthesia

Anaesthesia recommendations for patients suffering from

Allgrove syndrome

Disease name: Allgrove Syndrome

ICD 10: E27.4

Synonyms: Triple A syndrome, 4 A Syndrome, Achalasia-Addisonianism-Alacrima syndrome

Disease summary: Allgrove Syndrome (AS) is rare autosomal recessive disorder characterised by achalasia cardia, alacrimia and adrenal insufficiency, which is generally adreno-corticotropic hormone (ACTH) resistant, and neurological abnormalties. Mutations have been identified in AAAS gene, located on chromosome 12q13 (type 2 keratin gene), that codes for ALADIN protein. The IVS14 and EVS9 are the most common mutations. Alacrimia is an early and pathognomic symptom, but achalasia (50 -100 %) and adrenal insufficiency (20 -54%) are the more common presenting features. Autonomic disturbances and other neurological symptoms (10- 23%) are rare. Patients may develop a variable combination of sensory-motor polyneuropathy amyotrophy, dysarthria, hyper-reflexia, muscle weakness, dementia, abnormal autonomic function, erectile dysfunction (adult) and intellectual impairment. Diagnosis is generally made in the first decade of life when they present with dysphagia, vomiting and failure to thrive due to achalasia, hyper-pigmentation of skin, shock due to adrenal insufficiency or seizures and coma due to severe hypoglycaemia. Typical dysmorphic facies including long thin face, long philthrum, narrow upper lip, down turned mouth and sparse eyelashes may also be seen. Keratitis punctata is the most common complication of alacrimia. Patients with adrenal insufficiency are generally on a maintainance dose of a glucocorticoid like hydrocortisone. Most patients with achalasia require frequent pneumatic dilatations or surgical interventions like Heller's myotomy.

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

Typical surgery

Heller's Myotomy for Achalasia Cardia (open or laparoscopic), baloon dilatation of esophagus under endoscopic control.

Type of anaesthesia

General anaesthesia with controlled airway in the form of an endo-tracheal tube is the standard practice.

Necessary additional diagnostic procedures (preoperative)

Barium swallow, esophageal manometry and upper GI endoscopy for achalasia cardia.
Cortisol levels and ACTH stimulation tests, serum electrolytes and serum glucose for adrenal insufficiency.

Schirmer's test for alacrimia.

Tests of autonomic dysfunction like heart rate and blood pressure response to standing, pilocarpine eye test and sweat test.

Motor or sensory impairment should be documented in consultation with a neurologist for medico-legal precautions.

Particular preparation for airway management

Patients are prone to recurrent respiratory tract infections due to regurgitation. Upper and lower respiratory tract symptoms must be ruled out. Active infections must be optimized. Surgery should be postponed if required.

Cuffed or micro cuffed Endo-tracheal tubes should ideally be used for airway protection and to prevent micro aspirations.

H_2 receptor blockers or proton pump inhibitors may be given for aspiration prophylaxis and for prevention of peptic ulcers due to stress dose of steroids.

Aspiration and decompression of stomach and esophageal contents by naso-gastric tube prior to induction.

Particular preparation for transfusion or administration of blood products

Maintenance intra-venous fluid, should be a dextrose containing fluid like 0.45% saline in 5% dextrose. Lactated ringer for intra-operative losses.

No contra-indications to blood transfusion, routine cross matching and transfusion related precautions should be taken.

Particular preparation for anticoagulation

No specific precautions or requirements mentioned in literature.

Particular precautions for positioning, transport or mobilisation

Patients are prone to autonomic disturbances. Positioning and transport should be slow and gradual. Laparoscopy may require head low position. Pneumo-peritoneum should be created slowly and intra abdominal pressure should be monitored properly.

Pressure points and bony prominences should be properly padded, especially in cases of sensory neuropathies and longer duration surgery. Similar precautions should be followed during transport.

Eyes should be properly lubricated and covered.

Probable interaction between anaesthetic agents and patients' long-term medication

Patients with adrenal insufficiency are on maintenance steroid doses, Cushing syndrome may be seen as a side effect. Stress doses of steroid are required peri-operatively and hydrocortisone is the drug of choice. The dose of hydocortisone and other steroids varies among different pediatric age groups and body weight [24,25].

Avoid etomidate as it is shown to depress adrenal function for 3-6 hours post administration. No direct interaction with other anaesthetic agents is mentioned in literature.

Anaesthetic procedure

General anaesthesia with airway being secured with endo-tracheal tube, preferably cuffed or micro-cuffed.

Induction should be done slowly, giving drug in aliquots, to prevent sudden cardiovascular collapse or autonomic instability.

Rapid sequence induction using short onset drug like rocuronium to minimise aspiration. Caution in use of succinyl choline, in presence of myopathies causing up-regulation of extra-junctional acetyl choline receptors (hyperkalemic response). Non depolarising blockers also may have a varying response in such cases. Should titrate dosing according to neuromuscular monitoring.

Maintainance of euglycaemia intra-operatively by use of Insulin infusion if required. Sometimes steroid infusion may also be required.

Particular or additional monitoring

Intraoperative monitoring of serum glucose, electrolytes (sodium, potassium).

Invasive blood pressure monitoring for early detection of any autonomic or hemodynamic disturbances.

Use of neuromuscular monitoring to titrate optimal dosing of neuro-muscular blocking drugs, and to optimize reversal and adequate recovery.

Intra-abdominal pressure monitoring, peak airway pressure monitoring, urine output and end tidal CO_2 to detect complications of pneumo-peritoneum.

Possible complications

Adrenal crisis leading to hypotension or shock, hypoglycaemia, hyponatremia or hyperkalemia (due to stress of surgery, infection or trauma).
Hyperglycemia due to steroids.

Post-operative care

Head end elevation and aspiration prophylaxis. Topical lubricating ointment for eyes.

Steroids should be tapered to maintenance dosing. Pain relief with intra venous paracetamol or per-rectal suppository.

Information about emergency-like situations/ Differential diagnostics

Adrenal crisis, hypotension and shock, should be differentiated from adverse effects or side effects of anaesthetic agents, or haemodynamic effects of autonomic dysfunction.

Hyploglycemia coma, hyponatremia may be a cause of delayed recovery from anaesthesia. Hyperkalemia may lead to dangerous arrhythmias or even cardiac arrest.

Ambulatory anaesthesia

May be practiced for shorter recurrent procedures like esophageal balloon dilatations.

Obstetrical anaesthesia

Not much literature on anaesthesia in this group of patients. Patients mostly encountered in the paediatric age group.

Literature and internet links

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