orphanaesthesia

Anesthesia recommendations for patients suffering from

Alpha-mannosidosis

Disease name: Alpha-mannosidosis

ICD 10: E77.1

Synonyms: Lysosomal alpha-D-mannosidase deficiency, Alpha-Mannosidase B Deficiency

Alpha-mannosidosis is caused by the lack of the lysosomal enzyme alpha-mannosidase due to a mutation in MAN2B1, located on chromosome 19. Lack of alpha-mannosidase causes disturbed glycoprotein catabolism. This causes an excessively high level of mannose-rich oligosaccharides in many tissues. It is a progressive disease with mental retardation, skeletal and muscle abnormalities, recurrent infections, psychiatric symptoms, and compromised pulmonary function. Symptoms progress slowly over decades and long-term prognosis is poor. Prevalence is estimated to be from 1 per 300 000 to 1 per 1 000 000.

Almost all patients suffer from hearing loss and a high incidence of accompanying autoimmune disorders (e.g. lupus erythemadoses, pancytopenia, hypothyroidism or primary biliary cirrhosis) is reported.

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

Muscle biopsies, insertion of central venous catheters and Port-A-Caths; general surgery; imagining studies (e.g. CT and MRI).

Type of anaesthesia

There is no definite recommendation for either general or regional anaesthesia.

However, many of the patients will be children or adolescents and will not be able to cope with the use of regional anaesthesia as sole anaesthetic method.

Anaesthesia can be performed as total intravenous anaesthesia as well as with the use of volatile anaesthetics without reported complications.

Necessary additional diagnostic procedures (preoperative)

A lung function test should be considered preoperatively if the patient lung function seems affected and the urgency of the operation permits further testing.

Particular preparation for airway management

These patients have a potentially difficult airway and high risk of upper airway obstruction; causes may include accumulations in mucous membranes, short and stiff neck with cervical instability, small foramen magnum and dependency of high muscle tone to keep airways open.

This could potentially worsen with the progress of the disease so that adult patients are more a risk of a difficult airway; however data on this are missing.

One case series (ref 2) showed that not all patients with alpha-mannisodosis will have a difficult airway. Assumptions about the difficulty of the airway should therefore not be based soly of the diagnosis of alpha-mannosidosis. Instead, each patient should undergo an individual preoperative airway assesement.

Fiberoptic intubation does not have to be first choice unless an individual assessment of the airways indicates so. Nonetheless, equipment for fiberoptic intubation should always be prepared and readily when given anaesthesia all patients with Alpha-mannosidosis.

Particular preparation for transfusion or administration of blood products

Standard precautions

Particular preparation for anticoagulation

Standard precautions

Particular precautions for positioning, transport or mobilisation

Standard precautions

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

Standard precautions

Anaesthesiologic procedure

Oral midazolam as premedication should be considered prior to procedures in anxious patients.

A higher risk of aspiration may be considered.

Due to the risk of a difficult airway, anaesthesia with maintenance of spontaneous breathing should be considered as first choice if the nature of the procedure permits.

In a case series of 14 anaesthesias (ref 2), there were no cases of difficult bag-mask ventilation and no cases of significant difficulties with intubation. The patients in the series were aged 7–17 years) and weighed a median of 47.5 kg (range 18.6–68.3 kg); it should be noted that these patients were a selected group participating in a clinical trial.

There are reports or personal communications/experiences about the use of several anaesthetics (thiopental, propofol, ketamine), volatile anaesthetics (sevoflurane, nitrous oxide), opioids (fentanyl, alfentanil, remifentanil. Morphine) and non-depolarizing muscle relaxants (cisatracurium, rocuronium, vecuronium, mivacurium) (ref 5).

If succinylcholine can be used safely is without evidence. But assuming myopathic signs it should be avoided.

The use of sugammadex for reversal of neuromuscular blockade is not reported.

Particular or additional monitoring

Standard monitoring according to common guidelines should be appropriate.

Possible complications

In a case series (see references) postoperative nausea and/or vomiting were noted after 5 of the 14 anaesthesia sessions. Thus, prophylaxis for postoperative nausea and vomiting should be considered.

Postoperative care

All patients should be carefully observed postoperatively, e.g., for airway edema (which in comparable disorders poses a high risk).

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.:*

No data available.

Ambulatory anaesthesia

No data available.

Obstetrical anaesthesia

No data available.

Literature and internet-links

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