

orphananesthesia

Anaesthesia recommendations for patients suffering from **Central Core Disease**

Disease name: Central core disease

ICD 10: G71.2

Synonyms: Shy-McGee syndrome

Central core disease (CCD) is an inherited (mostly dominant) neuromuscular disorder characterised by central cores in type I fibers on muscle biopsy and clinical features of a congenital myopathy. Prevalence is unknown because of variable expression and incomplete penetrance. Associated in approximately 25% of cases with malignant hyperthermia because of gene proximity or overlap: the mutation then involves gene RYR1 (19q13.1-13.2). In case of rare recessive transmission gene MYH7 (14q11.1) is involved. Multimicore, minicore myopathy and core-rod myopathy are closely related to central core disease and probably carry the same risk for malignant hyperthermia.

CCD typically presents in infancy with hypotonia and motor developmental delay and is characterized by predominantly proximal weakness, pronounced in the hip girdle.

Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

Typical surgery

Muscle biopsy; orthopedic surgery: correction of talipes equinovarus, scoliosis or dislocation of the hip or the patella.

Type of anaesthesia

Succinylcholine and volatile anaesthetics have to be strictly avoided.

General anaesthesia performed as total intravenous anaesthesia or regional anaesthesia can be done without complications equally. There are reports about the uneventful performance of spinal as well as epidural anaesthesia or a combination of both.

There is no contraindication for (analgo-) sedation beside common restrictions.

Necessary additional diagnostic procedures (preoperative)

Cardiac function is not typically impaired in patients with CCD, so preoperative cardiac function tests are not obligatory. There are some spare reports of cardiac involvement. In this case and in case of severe scoliosis preoperative echocardiography is necessary.

In some cases of recessive inheritance with neonatal onset respiratory system is impaired, and lung function tests maybe considered. This is also recommended in case of severe scoliosis.

If muscular weakness is present and regional anaesthesia is planned neurological consultation is helpful for juridical reasons.

Determination of preoperative creatine kinase level is not mandatory, but may be helpful in case of perioperative complications (e.g. rhabdomyolysis or malignant hyperthermia).

Particular preparation for airway management

Not reported.

Particular preparation for transfusion or administration of blood products

There is one small study showing a higher intraoperative blood loss during surgery for scoliosis in patients with neuromuscular diseases in general compared to idiopathic scoliosis. In CCD there is no evidence about bleeding abnormalities.

Particular preparation for anticoagulation

Not reported.

Particular precautions for positioning, transport or mobilisation

Not reported.

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

Not reported.

Anaesthesiologic procedure

Strictly avoid succinylcholine and any volatile anaesthetic because of the risk for malignant hyperthermia.

Use of opiates (remifentanyl, alfentanil, fentanyl, morphine), intravenous anaesthetics (propofol, midazolam), nitrous oxide, local anaesthetics (ropivacaine, bupivacaine) and non-depolarizing muscle relaxants (rocuronium, pancuronium) has been reported without complications.

When using non-depolarizing muscle relaxants no prolonged neuromuscular blockade is reported. Antagonisation of neuromuscular blockade with neostigmine was reported uneventful.

There is no need for prophylactic postoperative ventilation.

Particular or additional monitoring

Monitoring of the neuromuscular blockade is recommended.

Possible complications

All CCD patients are highly susceptible for malignant hyperthermia.

Also there are no reports about prolonged effects of non-depolarizing muscle relaxants it cannot be excluded. Therefore monitoring of the neuromuscular blockade is recommended.

Postoperative care

Avoid prolonged immobilization. Accompanying muscular atrophy may worsen disease.

Degree of postoperative supervision is depending on surgical procedure and preoperative condition of the patient. Intensive care is not mandatory.

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 disease

Disease triggered emergency-like situations are not common in CCD.

Respiratory function can be more likely impaired in case of acute respiratory tract infection.

Ambulatory anaesthesia

In cases of stable disease without respiratory impairment, ambulatory anaesthesia is possible according to common guidelines.

Obstetrical anaesthesia

Obstetrical anaesthesia can be done as general (without succinylcholine and volatiles) as well as regional anaesthesia. Be aware that in most neuromuscular disorders disease progression can occur during pregnancy.

Use of syntocinon was reported without complications.

After the use of dantrolene (treatment in case of malignant hyperthermia) uterine atony is reported.

Because of inheritance of the disease there is the possibility of impaired newborns with muscular hypotonia or respiratory distress.

Literature and internet links

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