orphanaesthesia

Anaesthesia recommendations for

Duchenne muscular dystrophy

Disease name: Duchenne muscular dystrophy

ICD 10: G71.2

Synonyms: Dystrophinopathy

Disease summary: Duchenne muscular dystrophy (DMD) is the most common and severe muscular dystrophy with an incidence of 1 in 3000 male newborns; it is caused by a mutation in the dystrophin gene located on chromosome Xp21. As de-novo mutations are frequent, a positive familial history is lacking in 30% of cases: in those cases, the mean age of diagnosis is between 3 and 5 years. Some females carrying the gene on one X can present with a muscular or cardiac pathology. This mutation results in a deficit of dystrophin, an important sarcolemmnal structural protein in muscle cells. The clinical course of DMD is severe and there is no causative therapy available but some patients are on chronic corticoid therapy because this slows the progression of the disease. The disorder is characterized by progressive skeletal muscle weakness with an early onset in childhood. Muscle remodelling with fibrous and fatty tissue leads to loss of ambulation at a mean age of 10 years. Most of these patients require corrective orthopaedic surgery in the early stage of the disease for foot deformities and later for severe scoliosis to improve quality of life. The main anaesthetic concern in the treatment of patients with DMD is the use of depolarizing relaxants and volatile (halogenated) anaesthetics because of the potential for severe hyperkalimia and rhabdomyolysis.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

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Typical surgery

Muscle biopsy; orthopaedic surgery, tendon releases, tendon transfers, correction of scoliosis.

Type of anaesthesia

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

Regional or local anaesthesia can be provided. There are reports of spinal, epidural and caudal anaesthesia without any complication. Ultrasound guidance is advisable, since landmarks may be obscured due to musculo-skeletal deformations.

Succinylcholine and volatile anaesthetics are best avoided because there is a risk of severe hyperkalemia and severe rhabdomyolysis. General anaesthesia is best performed as total intravenous anaesthesia. However, some authors have suggested that in special circumstances (eg difficult venous access), a short-lasting use of inhalation anaesthesia is acceptable as long as the anaesthesiologist is prepared to treat acute rhabdomyolysis). There is no risk of malignant hyperthermia.

Nitrous oxide can be used, but should be avoided in case of manifest cardiac involvement.

In young patients with an early stage of the disease (no cardiopulmonary involvement, ability to walk) there is no contraindication for (analgo-)sedation. In patients with advanced stage of the disease (cardiopulmonary involvement, pharyngeal muscle weakness, loss of ambulation) the performance of (analgo-)sedation should only be done after carefully calculating the individual risks, especially with respect to respiratory failure and risk of aspiration.

Necessary additional pre-operative testing (beside standard care)

DMD is a progressive muscular dystrophy and therefore patients with early stage of the disease have no relevant involvement of other organ systems beside the weakness of the skeletal muscle, whereas patients with advanced stage of the disease show severe involvement of the cardiac and pulmonary system. Therefore, with ongoing disease, cardiopulmonary testing is necessary.

Cardiac function tests including electrocardiography and echocardiography should be performed for evaluating presence of cardiomyopathy.

Lung function tests including lung volumes and blood gas analysis should be done to evaluate grade of pulmonary involvement. Recognize that there is no clear correlation between lung function and postoperative respiratory complications.

Creatine kinase levels are usually elevated but show no correlation with disease severity. Determination of a preoperative baseline is useful, if only to obtain a baseline level in case of perioperative complications like rhabdomyolysis. In a later disease stage, e.g. in wheelchair-bound patients, CK levels may return to normal levels.

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

Particular preparation for airway management

Own retrospective data showed a difficult intubation in 8 out of 219 patients (mask ventilation without problems). Although this is only weak evidence, the possibility of a difficult airway must be considered. Macroglossia is frequent.

Patients with advanced stage of disease present with weakness of the oropharyngeal muscles including swallowing difficulties and possible elevated risk of aspiration.

Particular preparation for transfusion or administration of blood products

There may be a higher requirement for blood products in patients with DMD during highly invasive surgery. There is some evidence for impaired platelet function, altered coagulation and fibrinolysis and decreased vessel reactivity. This may have clinical consequences. One small study showed a higher intraoperative blood loss during surgery for scoliosis in patients with neuromuscular diseases (role of osteoporosis?), especially with DMD compared to patients with idiopathic scoliosis.

Particular preparation for anticoagulation

There is no evidence to support the need of particular anticoagulation. But the impaired mobility of advanced stage patients may suggest a higher risk of postoperative thrombosis.

Particular precautions for positioning, transportation and mobilisation

Not reported.

Interactions of chronic disease and anaesthesia medications

Not reported. Provide steroid substitution in case of corticoid therapy.

Anaesthetic procedure

Avoid succinylcholine and any volatile anaesthetic (including washout of the anaesthesia machine before induction) because of the risk of hyperkalemic cardiac arrest and rhabdomyolysis.

In case of present cardiomyopathy, avoid nitrous oxide because of cardio-depressant effects.

Opiates, propofol, dexmedetomidine, ketamine and local anaesthetics have been used without any complication. Patients may require a higher dose of propofol or opiates.

Non-depolarizing neuromuscular blocking agents can be used safely in these patients, but show markedly differences in onset and duration of action. In general, onset of

neuromuscular block is delayed and duration is markedly prolonged. These effects are pronounced in advanced disease and with rocuronium compared to mivacurium.

Antagonisation of neuromuscular blockade with pyridostigmine or neostigmine seems to be possible. In case of doubt, prefer ventilation until spontaneous recovery. There are reports about the successful administration of sugammadex.

There is one report about higher toxicity of paracetamol in patients with DMD.

Particular or additional monitoring

Monitoring of the neuromuscular blockade is strictly recommended if any neuromuscular blocking agent is used: it is useful to obtain baseline values before injection of the non-depolarizing neuromuscular blocking agent.

Monitor body temperature to avoid shivering and increased oxygen demand.

In case of high-risk surgery, major fluid shifts or advanced disease, arterial cannulation for invasive blood pressure measurement and central venous line placement is recommended. In case of cardiomyopathy, transoesophageal echocardiography is very useful.

Possible complications

Patients with DMD are at risk for hyperkalaemic cardiac arrest (succinylcholine) and rhabdomyolysis (volatile anaesthetics).

Sedative drugs (benzodiazepines) can cause respiratory insufficiency.

Muscle relaxants show up to a 4 times prolongation of neuromuscular block. This effect is dependent on the stage of the disease.

DMD patients are at risk for respiratory and cardiac insufficiency.

Post-operative care

The degree of postoperative monitoring and a possible admission to intensive care are depending on both surgical procedure and preoperative condition of the patient.

Avoid prolonged immobilization. Accompanying muscular atrophy may worsen disease.

In case of necessary postoperative ventilation, strive for early weaning (e.g., non-invasive ventilation), avoid prolonged ventilation.

Disease-related acute problems and effect on anaesthesia and recovery

As mentioned above.

Ambulatory anaesthesia

Ambulatory anaesthesia (according to common guidelines), if at all, should only be done in DMD patients with early disease (no cardiopulmonary symptoms) and low risk surgery.

Obstetrical anaesthesia

Females suffering from DMD are a real rarity, due to spontaneous mutation in a carrier of DMD. There are no reliable data available.

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