orphananesthesia

# Anaesthesia recommendations for patients suffering from

# Duchenne muscular dystrophy

**Disease name:** Duchenne muscular dystrophy

**ICD 10:** G71.2

**Synonyms:** Dystrophinopathy

Duchenne muscular dystrophy (DMD) is the most common and severe muscular dystrophy with an incidence of 1 in 3,000 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 sarcolemmal 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 corticotherapy because this slows the progression of the disease.This disorder is characterized by progressive skeletal muscle weakness with an early onset in childhood. Muscle reorganization with fatty infiltration and increase in fibrous tissue leads to loss of ambulation by the age of 10 yr. Most of these patients require corrective orthopedic 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 halogenated agents because of the potential for hyperkalemic cardiac arrest and rhabdomyolysis.

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong



© Anästh Intensivmed 2014;55:S544-S561 Aktiv Druck & Verlag GmbH

## 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.

Succinylcholine and volatile anaesthetics have best avoided because there is a risk of hyperkalemic cardiac arrest or severe rhabdomyolysis. There is no risk of malignant hyperthermia: some authors therefore agree that in special circumstances (e.g. difficult venous access) a short-lasting use of inhalation anaesthesia is possible as long as the anaesthesiologist is prepared to treat acute rhabdomyolysis).

General anaesthesia has to be performed as total intravenous anaesthesia. Nitrous oxide can be used, but should be avoided in case of manifest cardiac involvement.

Regional or local anaesthesia can be done. There are reports of spinal, epidural and caudal anaesthesia without any complication.

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 diagnostic procedures (preoperative)

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 test including electrocardiography and echocardiography should be performed for evaluating presence of cardiomyopathy.

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

Creatine Kinase Level is usually very elevated (and can be used as a screening tool) but shows no correlation with disease severity. Determination of preoperative baseline is useful if only to obtain a baseline level in case of perioperative complications like rhabdomyolysis. In chronic disease stage, e.g. in wheelchair-bound patients, CK levels may turn 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 possibility of difficult airway must be taken into account. 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 high invasive surgery. There is some evidence for deficiency of platelet function, altered coagulation and fibrinolysis and impaired 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, transport or mobilisation

Not reported.

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

Not reported. Provide steroid substitution in case of corticotherapy.

## Anaesthesiologic 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 and local anaesthetics have been used without any complication. Patients may require a higher dose of propofol or opiates.



© Anästh Intensivmed 2014;55:S544-S561 Aktiv Druck & Verlag GmbH

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 doubt prefer ventilation until spontaneous recovery. There is one single report about the successful administration of sugammadex.

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

There is no need for prophylactic postoperative ventilation if neuromuscular blockade was monitored consequently.

## 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 fluids shifts or advanced disease arterial cannulation for invasive blood pressure measurement and central venous line placement is recommended. In case of cardiomyopathy, transesophageal 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.

## Postoperative care

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

Avoid prolonged immobilization. Accompanying muscular atrophy may worsen disease.

In case of necessary postoperative ventilation intend for aggressive weaning (e.g. non invasive ventilation), avoid prolonged ventilation.



© Anästh Intensivmed 2014;55:S544-S561 Aktiv Druck & Verlag GmbH

## 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 DMD.

## 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.

**Literature and internet links**

1. Almenrader N, Patel D. Spinal fusion surgery in children with non-idiopathic scoliosis: is there a need for routine postoperative ventilation? Br J Anaesth 2006;97(6):851-857
2. Barohn RJ, Levine EJ, Olson JO, Mendell JR. Gastric hypomotility in Duchenne's muscular dystrophy. N Engl J Med 1988;319(1):15-18
3. Birnkrant DJ, Panitch HB, Benditt JO, Boitano LJ, Carter ER, Cwik VA, et al. American College of Chest Physicians consensus statement on the respiratory and related management of patients with Duchenne muscular dystrophy undergoing anesthesia or sedation. Chest 2007;132(6):1977-1986
4. Breucking E, Reimnitz P, Schara U, Mortier W. [Anesthetic complications. The incidence of severe anesthetic complications in patients and families with progressive muscular dystrophy of the Duchenne and Becker types]. Anaesthesist 2000;49(3):187-195
5. Bushby KM, Goodship JA, Nicholson LV, Johnson MA, Haggerty ID, Gardner-Medwin D. Variability in clinical, genetic and protein abnormalities in manifesting carriers of Duchenne and Becker muscular dystrophy. Neuromuscul Disord 1993;3(1):57-64
6. Bushby KM, Hill A, Steele JG. Failure of early diagnosis in symptomatic Duchenne muscular dystrophy. Lancet 1999;353(9152):557-558
7. Caliskan E, Sener M, Kocum A, Aribogan A. Duchenne muscular dystrophy: how I do it? Regional or general anesthesia? Paediatr Anaesth 2009;19(6):624-625
8. Ciafaloni E, Fox DJ, Pandya S, et al. Delayed diagnosis in Duchenne muscular dystrophy: data from the Muscular Dystrophy surveillance, tracking and reserach network. J Pediatr 2009; 155: 380-5
9. Coral-Vazquez R, Cohn RD, Moore SA, Hill JA, Weiss RM, Davisson RL, et al. Disruption of the sarcoglycan-sarcospan complex in vascular smooth muscle: a novel mechanism for cardiomyopathy and muscular dystrophy. Cell 1999;98(4):465-474
10. De Boer H, Van Esmond J, Booij LHJD, Driessen JJ. Reversal of rocuronium-induced profound neuromuscular block by sugammadex in Duchenne muscular dystrophy. Paediatr Anaesth 2009; 19(12):1226-8
11. English KM, Gibbs JL. Cardiac monitoring and treatment for children and adolescents with neuromuscular disorders. Dev Med Child Neurol 2006;48(3):231-235
12. Fairfield MC. Increased propofol requirements in a child with Duchenne muscular dystrophy.
13. Anaesthesia 1993;48(11):1013
14. Farah MG, Evans EB, Vignos PJ, Jr. Echocardiographic evaluation of left ventricular function in Duchenne's muscular dystrophy. Am J Med 1980;69(2):248-254
15. Feldman S, Karalliedde L. Drug interactions with neuromuscular blockers. Drug Saf 1996;15(4):261-273
16. Finsterer J, Stollberger C. Cardiac involvement in primary myopathies. Cardiology 2000;94(1):1-11. 16.Forst J, Forst R, Leithe H, Maurin N. Platelet function deficiency in Duchenne muscular dystrophy. Neuromuscul Disord 1998;8(1):46-49
17. Girshin M, Mukherjee J, Clowney R, Singer LP, Wasnick J. The postoperative cardiovascular arrest of a 5-year-old male: an initial presentation of Duchenne's muscular dystrophy. Paediatr Anaesth 2006;16(2):170-173
18. Harper CM, Ambler G, Edge G. The prognostic value of pre-operative predicted forced vital capacity in corrective spinal surgery for Duchenne's muscular dystrophy. Anaesthesia 2004;59(12):1160-1162
19. Hayes J, Veyckemans F, Bissonnette B. Duchenne muscular dystrophy: an old anesthesia problem revisited. Paediatr Anaesth 2008;18(2):100-106
20. Hayes J Gurnaney H, Brown A, Litman RS. Malignant hyperthermia and muscular dystrophies.Anesth Analg 2009; 109: 1043-8
21. Gurnaney H, Brown A, Litman RS. Malignant hyperthermia and muscular dystrophies. Anesth Analg 2009; 109: 1043-8)
22. Milne B, Rosales JK. Anaesthetic considerations in patients with muscular dystrophy undergoing spinal fusion and Harrington rod insertion. Can Anaesth Soc J 1982;29(3):250-254
23. Muenster T, Schmidt J, Wick S, Forst J, Schmitt HJ. Rocuronium 0.3 mg x kg-1 (ED95) induces a normal peak effect but an altered time course of neuromuscular block in patients with Duchenne's muscular dystrophy. Paediatr Anaesth 2006;16(8):840-845
24. O'Higashi T, Shirakami G, Sasai S, Shinomura T, Kato S, Tomoda K. [Spinal anesthesia for patients with progressive muscular dystrophy]. Masui 1995;44(5):723-728

25. Ross AK. Muscular dystrophy versus mitochondrial myopathy: the dilemna of the undiagnosed hypotonic child.(editorial). Pediatr Anesth 2007; 17: 1-6
26. Saito T, Takenaka M, Miyai I, Yamamoto Y, Matsumura T, Nozaki S, et al. Coagulation and fibrinolysis disorder in muscular dystrophy. Muscle Nerve 2001;24(3):399-402
27. Sax TW, Rosenbaum RB. Neuromuscular disorders in pregnancy. Muscle Nerve 2006;12:12
28. Schmidt J, Muenster T, Wick S, Forst J, Schmitt HJ. Onset and duration of mivacurium-induced
29. neuromuscular block in patients with Duchenne muscular dystrophy. Br J Anaesth 2005;95(6):769- 772
30. 29.Shapiro F, Sethna N. Blood loss in pediatric spine surgery. Eur Spine J 2004;13 Suppl 1:S6-17 30.Smith PE, Edwards RH, Calverley PM. Oxygen treatment of sleep hypoxaemia in Duchenne muscular dystrophy. Thorax 1989;44(12):997-1001
31. Turturro F, Rocca B, Gumina S, De Cristofaro R, Mangiola F, Maggiano N, et al. Impaired primary hemostasis with normal platelet function in Duchenne muscular dystrophy during highly-invasive spinal surgery. Neuromuscul Disord 2005;15(8):532-540
32. Wick S, Muenster T, Schmidt J, Forst J, Schmitt HJ. Onset and duration of rocuronium-induced neuromuscular blockade in patients with Duchenne muscular dystrophy. Anesthesiology 2005;102(5):915-919
33. Wollinsky KH, Weiss C, Gelowicz-Maurer M, Geiger P, Mehrkens HH, Naumann T. [Preoperative risk assessment of children with Duchenne muscular dystrophy and relevance for anesthesia and intra- and postoperative course]. Med Klin (Munich) 1996;91 Suppl 2:34-37.

**Last date of modification: June 2011**

---

*These guidelines have been prepared by:*
**Author**
**Tino Muenster**, Anaesthesiologist, University Hospital Erlangen, Germany
tino.muenster@kfa.imed.uni-erlangen.de

**Peer revision 1**
**Francis Veyckemans**, Anaesthesiologist, University Hospital St. Luc, Brussels, Belgium
francis.veyckemans@uclouvain.be

**Peer revision 2**
**Maggie Walter**, Neurologist, Friedrich-Baur-Institute, Ludwig-Maximilians-University of Munich, Germany
maggie.walter@lrz.uni-muenchen.de

**Benedikt Schosser**, Neurologist, Friedrich-Baur-Institute, Ludwig-Maximilians-University of Munich, Germany
benedikt.schosser@med.uni-muenchen.de

---



© Anästh Intensivmed 2014;55:S544-S561 Aktiv Druck & Verlag GmbH