Acute heart failure during spinal surgery in a boy with Duchenne muscular dystrophy

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Patients with Duchenne muscular dystrophy (DMD) are at high risk of perioperative complications. DMD may be accompanied by heart failure resulting from dystrophic involvement of the myocardium, which can be subclinical in the early stages of the disease. This case demonstrates that a normal preoperative ECG and echocardiograph cannot exclude the development of heart failure during anaesthesia in DMD patients undergoing major surgery.

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Duchenne muscular dystrophy (DMD) is a sex-linked muscular dystrophy accompanied by almost complete loss of the muscular protein dystrophin, leading to a weakened sarcolemma. The disease is characterized by severe proximal muscle weakness, progressive degeneration, and fatty infiltration of the muscles. The symptoms appear at the age of 2–6 yr, and patients are usually wheelchair-bound by 10 yr. The progressive nature of the disorder results in restrictive pulmonary disease, multiple contractures and scoliosis.

Patients with DMD are considered to be at high risk of perioperative complications. These may be related to the administration of succinylcholine or halothane, as the disease is associated with malignant hyperthermia.1–4 But DMD is also accompanied by heart failure attributable to dystrophic involvement of the myocardium.5 The cardiomyopathy of DMD is characterized by fibrosis of the posterobasal and contiguous lateral wall of the left ventricle, and is associated with arrhythmia, ventricular dilatation and cardiac insufficiency.6 Typical abnormal electrocardiographic patterns are tall R waves in the right chest leads, deep Q waves in the left precordial leads, biventricular hypertrophy and sinus tachycardia. In addition to using ‘trigger-free’ anaesthesia and testing lung function preoperatively, echocardiography is therefore recommended for patients with DMD.7 We present a case report which demonstrates, however, that normal preoperative echocardiography and electrocardiography cannot exclude the development of heart failure in patients with DMD during anaesthesia for major surgery.
Case report

An 11-yr-old boy, 140 cm tall, weighing 40 kg, ASA physical status class III, was admitted for elective thoracolumbar stabilization. His medical history was dominated by DMD diagnosed by histological examination and molecular genetic investigations. Since the age of 9 yr he had been wheelchair-bound. Preoperatively his lung function showed a restrictive pattern with a vital capacity (VC) of 0.8 l (33% of normal), but the chest X-ray and electrocardiogram were normal apart from a scoliosis. Preoperative transthoracic echocardiography (TTE) showed a normal cardiac anatomy and good ventricular function with a fractional shortening (FS) of 44% (Fig. 1A). Physical examination was unremarkable, and the results of laboratory tests were all within normal limits, except a plasma creatine kinase (CK) of 1483 U litre\(^{-1}\).

After fasting for 6 h and premedication with midazolam 7.5 mg p.o. 30 min preoperatively, anaesthesia was induced with sufentanil and propofol and maintained with propofol 4–10 mg kg\(^{-1}\) h\(^{-1}\) and repeated doses of sufentanil. Rocuronium bromide was used to facilitate orotracheal intubation. A central venous catheter, an invasive arterial blood pressure catheter and a urine catheter were inserted. Surgery was performed in the prone position. The anaesthetic was uneventful until 4 h after induction. Blood loss was corrected by transfusion of cell-saver solution 350 ml and packed blood cells 250 ml. Haemoglobin was maintained at 9–11 g dl\(^{-1}\). Suddenly, without obvious reason, an episode of hypotension (mean arterial pressure fell from 90 to 30–40 mm Hg) and tachyarrhythmia (up to 200 beats min\(^{-1}\)) were noted. Hypovolaemia, pneumothorax, pericardial tamponade, anaphylaxis and malignant hyperthermia were excluded by laboratory, intraoperative x-ray and clinical findings (Fig. 2). We confirmed by x-ray that the tip of the central venous catheter was outside the right atrium and thus was not the cause of the arrhythmia. The episode was controlled by administration of epinephrine 200 \(\mu\)g and norepinephrine 20 \(\mu\)g min\(^{-1}\) over 10 min. Haemodynamic values were stable within 60 min (Fig. 2).

After the end of surgery and on the return of the patient to the supine position, a second episode of hypotension and tachyarrhythmia without signs of acute bleeding, metabolic derangement or any other cause occurred. Only by administration of catecholamines (epinephrine 200 \(\mu\)g and norepinephrine 400 \(\mu\)g) could the patient be stabilized. An echocardiograph was performed which showed poor biventricular function with left ventricular fractional shortening of 13% (Fig. 1) and insufficiency of both atrioventricular valves (grade II). After haemodynamic stabilization, the patient was transferred to the intensive care unit and therapy was optimized with epinephrine 3 \(\mu\)g min\(^{-1}\) and milrinone. The tachyarrhythmia was treated with amiodarone. A diuresis was induced with furosemide. Sixteen hours later, after stabilization of the patient’s...
haemodynamics, he was extubated without any complications. After stopping the milrinone, 5 days after surgery, a third episode of hypotension occurred. Echocardiography again showed poor left ventricular function (fractional shortening 13%). Therapy was optimized with enalapril and digoxin. A further echocardiograph 10 days after the operation showed recovery of left ventricular function (fractional shortening 41%), so the patient was allowed to leave the intensive care unit (Fig. 1C).

Discussion

The episodes of hypotension and tachycardia during spinal surgery in this young boy with Duchenne muscular

Fig 2 Patient data. Changes in oxygen saturation (SaO₂), end-expiratory carbon dioxide (EECO₂), central venous pressure (CVP), heart rate (HR), blood pressure (BP, systolic, diastolic and mean), temperature (Temp), inspired oxygen concentration (FiO₂) and laboratory findings for the interval before (BL) and 30 min after induction of anaesthesia (30 min ai), immediately before (pre 1) the first hypotension event (1) and immediately before the second (pre 2) hypotension event (2). Grey boxes mark the two events of hypotension and tachycardia.
Duchenne dystrophy and acute heart failure

dystrophy were probably due to acute cardiomyopathy. It is well known that dystrophic involvement of the myocardium does occur in young patients with DMD. The weakened heart may have been unable to cope with the transfused blood, colloid and crystalloid solutions during the surgical procedure consequent to the blood loss, and to the changes in volume load after turning the patient back to the supine position. Interestingly, the preoperative echocardiogram showed good left ventricular function and did not suggest underlying heart failure. Thus, preoperative echocardiography may not reflect in each patient an intraoperative ability to respond adequately to myocardial stress. It has to be questioned whether preoperative transthoracic echocardiography and transoesophageal echocardiography are adequate indicators of myocardial reserve in this group of patients.

Several complications in association with general anaesthesia have been reported in the last two decades in patients with DMD. Larsen and colleagues investigated the frequency and severity of anaesthetic complications in a retrospective study of 84 operations in patients with DMD. No case presenting with acute heart failure was noted. The complications were almost exclusively related to the use of succinylcholine and to malignant hyperthermia. However, in 70.5% of the patients an abnormal electrocardiogram was observed. In a recent German study, 444 cases of dystrophic involvement of the myocardium were observed. In a recent German study, 444 cases of dystrophic involvement of the myocardium were observed.

Several complications, including cardiac arrest, occurred in young children with previously undiagnosed muscular dystrophy who received inhalational agents or succinylcholine. Anaesthetic risks and complications in 101 patients with DMD were investigated by Wollinsky and colleagues. Several complications could have been prevented by avoiding anaesthetic agents with a high trigger potential for malignant hyperthermia. The risk of cardiac insufficiency or arrhythmia was more pronounced in the older patients and could have been predicted by the findings in the preoperative echocardiograph. No patient with a normal preoperative echocardiograph developed heart failure perioperatively.

In contrast, Sethna and colleagues pointed out that there is no variable which preoperatively identifies patients who will develop complications during anaesthesia. He reported a 13-year-old boy scheduled for scoliosis repair. The preoperative echocardiograph showed normal left ventricular size and function. The patient suddenly developed hypotension and bradycardia intraoperatively, which was unresponsive to catecholamines and cardiopulmonary resuscitation. Autopsy showed cardiomyopathy with dilatation, hypertrophy and fibrosis. This case is in concordance with our findings that echocardiography at rest may not be sufficiently sensitive to estimate the intraoperative cardiac risk in patients with DMD. Sethna and colleagues suggested that several complications may be minimized by using narcotics instead of potent volatile anesthetics, and by using non-depolarizing muscle relaxants instead of succinylcholine. However, the present case report demonstrates that, by using propofol and sufentanil for anaesthesia, and rocuronium bromide for muscle paralysis, the perioperative management of patients with DMD can still cause major problems.

Propofol given by infusion may cause hypotension because of its negative inotropic properties. In the present case propofol was given in the usual dose range. Unfortunately, we do not know the propofol blood concentrations in this patient. It is unclear whether the continued administration of propofol by infusion resulted in overdosage. Routine intraoperative monitoring of brain electrical activity by, for instance, the electroencephalogram may have provided useful information in this respect.

In the last 2 yr there have been several reports about the ‘propofol-infusion syndrome’, defined as the constellation of otherwise unexplained myocardial failure, metabolic acidosis and rhabdomyolysis in the setting of prolonged, high-dose propofol infusion (>4.8–6 mg kg\(^{-1}\) h\(^{-1}\) for >48–72 h). However, a propofol-infusion syndrome-mediated circulatory insufficiency alone seems unlikely to be the cause of the problem in our patient as the hypotension and tachyarrhythmia occurred only 4 h after starting the propofol administration. But in this case we cannot exclude a contribution from the negative inotropic effects of propofol to the episode of severe heart failure.

Interestingly, female carriers of DMD can develop cardiomyopathy in the absence of skeletal muscle symptoms. Larach and colleagues pointed out that 48% of paediatric patients with cardiac arrest during anaesthesia had an unrecognized myopathy, and 67% of them were associated with hyperkalaemia. To decrease the incidence of myopathy-associated cardiac arrest during anaesthesia, screening of young male patients for occult myopathy has been recommended.

As preoperative echocardiography may not reflect the intraoperative ability of the diseased myocardium to respond to stress, a more sensitive method of assessing myocardial dysfunction in patients with DMD is needed. Angermann and colleagues proposed that stress echocardiography using angiotensin revealed a significantly reduced fractional shortening in physically disabled patients with DMD. They also demonstrated a correlation between clinical symptoms, abnormal echocardiographic findings and the extent of the skeletal muscle disease. Stress echocardiography detected latent heart failure in these patients and identified inducible contraction abnormalities in many patients with DMD. It might therefore be beneficial in detecting unrecognized cardiac problems preoperatively.

This case also raises the question of what intraoperative monitoring should be used for detection of cardiac abnormalities and for adjusting therapy. Most importantly, intraoperative transoesophageal echocardiography, pulmonary arterial catheterization and pulse contour analysis (PCCO) may be useful in this respect. Further studies are...
needed to investigate the importance of these tools, but transoesophageal echocardiography seems to be superior as it gives more detail of the anatomy (e.g. dilatation of the ventricle).

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