

Anesthesia in Children With Muscular Dystrophy: Important Information for Your Son's Doctor

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From Parent Project Newsletter 4/97

When a patient with muscular dystrophy is subjected to general anesthesia, a number of serious problems may arise. As a parent, it might be useful to be informed about these possible anesthesia-related risks and how these risks can be minimized by careful selection of the administered anesthetic agents. Therefore, a brief overview of the literature and some of the current ideas and concepts will be discussed.

Anesthesia-related complications in children with muscular dystrophy can be subdivided into general and specific categories.

General

Muscular dystrophy not only affects the muscles of the extremities, but as the disease progresses, heart and respiratory muscles become involved as well.

Almost all anesthetics decrease contractility of the heart. Many children with muscular dystrophy have an impaired cardiac function (cardiomyopathy). As a result, difficulties may arise with the circulation in the perioperative period.

Prolonged spontaneous respiration during general anesthesia may result in inadequate ventilation of the lungs with increased carbon dioxide levels in the blood. This in turn can cause severe heart rhythm disturbances, especially in those patients with an already impaired cardiac function and even more so in combination with certain (volatile) anesthetics. During the majority of general anesthetic procedures, however, the respiratory function is controlled by a mechanical respirator via insertion of a tube in the upper airways, which requires the use of drugs resulting in a temporary relaxation of the muscles. The administration of these muscle relaxants also carries specific risks which will be discussed below.

Following general anesthesia, problem may arise when the patient has to regain his respiratory function. The respiratory drive is diminished as a result of residual drug effects, coughing is impaired and aspiration easily occurs. All of this may result in low levels of oxygen and high levels of carbon dioxide in blood. Furthermore, certain areas of the lung may collapse (atelectasis) impairing oxygenation of the blood and increasing the risk of respiratory infections.

Therefore, it is important (and for large operations even mandatory) to perform extensive preoperative screening, not only to estimate the perioperative risks, but to provide optimal perioperative monitoring. The screening may include a cardiac ultrasound, an electrocardiogram, pulmonary functions tests as well as blood gases.

Specific

The membranes of muscle fibers and/or the receptors on the muscle cells can alter as a result of the disease, both in function as well as in number. Receptors are specific proteins on a cell membrane, which upon stimulation, modulate the function of the cell.

In patients who have been bedridden for a long period or who are unable to actively use their muscles, the administration of the "depolarizing" muscle relaxant succinylcholine may result in the release of large amounts of the ion potassium from the muscle cell into the blood stream, since

depolarizing muscle relaxants cause the muscles to contract briefly before they relax. This sudden increase in potassium concentration in the blood may result in life-threatening heart rhythm disturbances. The "non-polarizing" muscle relaxants (for instance vecuronium, atracurium or mivacurium) do not initiate muscle contraction and, therefore their use does not carry this risk. Nonetheless, the affected muscles may have become more sensitive to non-depolarizing muscle relaxants and, therefore, the dose administered has to be carefully chosen.

The brief but profound muscle contraction associated with the use of succinylcholine leads to elevated levels of creatinine kinase (CK) and myoglobin which can be detected in the blood afterwards. The use of succinylcholine and inhalational (volatile) anesthetics, such as halothane, can result in increased muscle breakdown, called rhabdomyolysis. Muscle break down leads to the release of high concentrations of potassium, CK and myoglobin in the circulation. As mentioned previously, high concentrations of potassium in the blood may result in cardiac arrest and the high concentrations of muscle-proteins may severely impair the kidney function.

In addition, the use of succinylcholine and inhalational anesthetics may result in a syndrome called malignant hyperthermia. Malignant hyperthermia is a life-threatening disturbance of the calcium-homeostasis in the muscle cell provoked by certain anesthetics. Succinylcholine and inhalational anesthetics are particularly renowned for causing this syndrome. Malignant hyperthermia occurs relatively frequent in patients with muscular dystrophy. Malignant hyperthermia is characterized by an extremely elevated metabolism within the muscle cell. As a result the temperature of the entire body rises to life-threatening levels, oxygen consumption and carbon dioxide production of the body increase dramatically and waste products are released into the circulation. It is of extreme importance that this syndrome is readily recognized so that the appropriate measures can be taken, including drastic cooling of the body and prompt initiation of therapy with the drug dantrolen. Unfortunately, despite appropriate measures, the course of this syndrome is often fatal. Children with muscular dystrophy should always be considered of being at high risk for the development of malignant hyperthermia and, therefore, the high risk anesthetics should be avoided.

Conclusions

General anesthesia is accompanied by a number of important risks. When general anesthesia is required in order to undergo a specific procedure succinylcholine and inhalational anesthetics need to be AVOIDED.

Painkillers (opioids), the anesthetic propofol, midazolam and hypnomidate can probably all be used safely. When it is essential to use muscle relaxants, short acting, non-depolarizing muscle relaxants can be used albeit at a reduced dose (one-fourth to one-fifth of the usual dose). Controversy exists as to whether antagonizing these muscle relaxants with cholinesterase inhibitors carry additional risks. Therefore, it is of utmost importance to inform your anesthetist as early as possible about the medical history of your child. The necessary pre-operative screening procedures (heart and lungs) can be performed and also the appropriate anesthetics can be selected. In addition, the anesthetist will be enabled to provide optimal peri-operative monitoring.

You may want to show this document to the anesthetist involved in caring for your child during any surgical or emergency procedure.

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