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Perioperative Care in Neuromuscular Scoliosis

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A 25-year-old male with Becker muscular dystrophy is referred from the neurosurgery clinic for preoperative evaluation for spinal stabilization. He has been hospitalized once for acute decompensated heart failure and has been treated for two episodes of pneumonia in the past 6 months. The patient and his family are concerned about the risk of heart failure and pulmonary complications.

Muscular dystrophy is a group of hereditary progressive diseases associated with cardiomyopathy, progressive decline in pulmonary function, and scoliosis, which are the major cause of morbidity and mortality. Muscle weakness, contractures, and progressive scoliosis impair pulmonary function, leading to hypoventilation and ineffective cough. Pulmonary function tests including arterial blood gas are essential to the evaluation of pulmonary reserve and degree of hypoventilation. Vital capacity of less than 35% predicted suggests that postoperative complications are likely and postoperative ventilatory support will be necessary. Preoperative physical therapy and breathing exercises may be beneficial in improving pulmonary reserve. Patients with hypoventilation may benefit from nocturnal ventilation started in the preoperative phase in consultation with a pulmonologist.

In a recent retrospective chart review, prior seizure history, unplanned blood loss, and unplanned staged surgery were factors statistically significantly associated with perioperative complications. Patients taking anticonvulsant drugs had significantly greater blood loss during anterior procedures. Cardiac evaluation per current guidelines and two-degree echocardiography is essential to quantify left ventricular ejection fraction. Succinylcholine administration is associated with life-threatening hyperkalemia and should be avoided in patients with muscular dystrophy. In the postoperative period, duration of intubation and the presence of atelectasis is a stronger predictor of infective complications. Prolonged prone positioning and hypotension can predispose to compartment syndrome, necessitating fasciotomy. The incidence of postoperative myocardial infarction is not well documented in this patient subgroup. Frank discussion with patients and their families regarding long-term use of artificial ventilation may be helpful to document patients' wishes. A thorough cardiac, pulmonary, and hematologic evaluation, along with awareness of specific adverse drug effects, will help guide care in the postoperative period. More studies need to be done to further evaluate the cardiac adverse events and protocols to better risk-quantify the same.