**Título:** ANAESTHETIC APPROACH TO A PATIENT WITH MCARDLE SYNDROME - A RARE METABOLIC DISEASE

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**Área Terapêutica/Tema:** Anestesia Regional (Regional Anaesthesia)

**Resumo:**

Introduction: McArdle disease or glycogen storage disease type V is a rare metabolic myopathy (1:100,000) characterized by glycogen deficiency in skeletal muscles (1). This disease has the potential for creating perioperative anaesthetic challenges, such as hypoglycaemia, rhabdomyolysis, myoglobinuria, acute renal failure and malignant hyperthermia (2). This is a case report about a patient with McArdle disease and his anaesthetic management.

Case report: A 63-year-old male, ASA II, BMI 30, with hypertension and diagnosed with McArdle disease submitted to an umbilical hernia repair. The patient described severe muscular pain and lack of strength during light exercise preventing an accurate evaluation of functional capacity. Patient had a Mallampati IV, chronic hoarseness and no neurological or kidney damage. We performed spinal anaesthesia with a 27G needle, 2.5 mcg sufentanil and 12.5 mg levobupivacaine. Active heating was maintained during the surgery and in the recovery room. The motor block was completely reversed after 4 hours with no alterations in muscle strength. Myoglobin and CK testing in the postoperatively were similar to the preoperative period. ICU care was considered although this wasn´t necessary as no complications were recorded perioperatively.

Discussion: Patients with McArdle disease are rare and rise several anaesthetic concerns. Using regional techniques, we avoid complications such as malignant hyperthermia, metabolic alterations and hypermetabolism. In addition, it allows the patient to adapt his positioning decreasing the risk of rhabdomyolysis. We should avoid shivering in these patients since muscle damage and increase in oxygen consumption can lead to important metabolic changes. Monitoring temperature and active patient warming during the surgery are also important measures.

Learning points: Patients with rare muscular diseases like McArdle syndrome are an anaesthetic challenge and demand a tailored anaesthesia. Regional techniques is the main option when the procedure allows it. Adequate monitoring and postoperative follow up also prevent complications. Choosing a spinal anaesthesia, we guaranteed the safety of our intervention without aggravating disease and without prolonging hospital stay or recovery.

References:

(1) BMC Anesthesiology. 2017; 17:134

(2) Acta Anaesthesiol Scand. 2005; 49: 1077–1083

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