**Título:** Anaesthetic Management of a Patient with Friedrich’s Ataxia

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**Área Terapêutica/Tema:** Prática baseada na evidência e melhoria da qualidade (Evidence-based Practice and Quality Improvement)

**Resumo:**

Friedrich’s Ataxia (FA) is a rare autosomal recessive disorder characterised by progressive gait and limb ataxia, dysarthria, lower limb areflexia, decreased position and vibration sense, muscular leg weakness and positive extensor plantar response, as well as non-neurological signs including hypertrophic cardiomyopathy and diabetes mellitus. There are few case reports in the literature of patients with FA undergoing general anaesthesia and their higher sensitivity to muscle relaxants makes the anaesthetic management challenging.

A 41-year-old woman, physical status ASA III, was scheduled for an elective modified radical mastectomy due to an invasive carcinoma with axillary lymph node metastasis. The patient had been diagnosed with FA 4 years before and presented with gait ataxia, dysarthria and lower limb areflexia. Aspiration prophylaxis was administered. An intravenous general anaesthesia was performed using a TCI of propofol. A second generation laryngeal mask was used. The procedure went by uneventfully and the patient was discharged home 2 days after surgery.

Anaesthetic considerations in FA patients should include: (1) avoidance of succinylcholine due to the risk of hyperkalaemia and mandatory monitoring of the neuromuscular blockade when using non-depolarizing muscle relaxants due to increased sensitivity, (2) considering rapid sequence induction and aspiration pneumonia prophylaxis due to the increased risk of aspiration, (3) avoidance of propofol-based total intravenous anaesthesia due to increased risk of propofol infusion syndrome, (4) favouring regional anaesthesia whenever possible and (5) prevention of possible peri or postoperative respiratory and cardiac complications. Even though some authors recommend avoiding propofol infusions, in this case we performed a general anaesthesia where propofol was safely used.

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