PO 12 - REGIONAL ANAESTHAESIA FOR AN ADULT PATIENT WITH WOLF-HIRSCHHORN SYNDROME – A CASE REPORT

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Background: Wolf-Hirschhorn syndrome (WHS) is a rare genetic disorder resulting from a partial deletion on the short arm of chromosome 4 (del(4p16.3)). Among other features, it is characterized by craniofacial abnormalities, mental retardation, muscle hypotonia, convulsions, scoliosis, kyphosis and talipes equinovarus. These abnormalities may present an anaesthetic challenge: possible difficulties in understanding and collaboration, airway management, unknown drug pharmacokinetics and pharmacodynamics, risk of seizures and malignant hyperthermia. Anaesthesia approach in WHS has been described in 9 articles since 1988: 3 malignant hyperthermia, 1 airway management and 5 anaesthetic considerations case reports.

Case Report: 21-year-old woman with WHS underwent surgical Achilles tendon elongation under spinal anaesthesia with levobupivacaine and sedation with ketamine and midazolam. Standard ASA monitoring was used. Multimodal analgesia (paracetamol 1g, metamizole 2g, ketorolac 30mg), and triple postoperative nausea and vomiting prophylaxis were given. Successful tendon elongation was performed, under an uneventful course of anaesthesia. The patient remained in the postanaesthesia care unit for 2 hours, with total motor block reversal confirmation. Hospital discharge occurred after 48 hours, without complications.

Discussion: Clinical evidence existing on general anaesthesia (GA) and WHS points out the risk of malignant hyperthermia and difficult airway. There are, at the moment, no spinal anaesthesia (SA) cases described in the literature – possible concerns are the lack of patient collaboration and anatomical difficulties. Considering the available options and its limitations, spinal anaesthesia in association with sedation was the chosen approach, with a successful outcome. SA could thus be a viable option for lower extremity surgery in WHS patients, not disregarding a careful airway evaluation and planned GA approach.

Learning Points: WHS patients may be a challenge for the anaesthesiologist due to its malformation spectrum and lack of reassuring literature. SA may be a successful and safe approach for WHS patients in need of lower extremity surgery.