

**P 2071****Safe anesthesia for imaging studies and diagnostic procedures in mucopolysaccharidosis: is it feasible?**

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Purpose: The high prevalence of airway obstruction and restrictive pulmonary disease in combination with cardiovascular manifestations poses a high anaesthetic risk to patients with mucopolysaccharidosis (MPS). The presence of macroglossia, tonsillar hypertrophy and swelling of tissues in laryngopharyngeal challenge the airway management. The anesthesiologist must be prepared to deal with inability to ventilate or intubate and also with sudden airway obstruction during anesthesia. On average 3 anesthetic surgical procedures will be performed in every MPS patient throughout his life. Our objective is to establish the use of appropriate anesthetic techniques especially in diagnostic tests performed outside the operating room. Methodology: All patients underwent a cerebrospinal fluid (CSF) flow and brain magnetic resonance imaging (MRI) followed by a standart lumbar puncture with the CSF opening pressure assessment in the radiology unit. Patients were assessed before the procedure and the anesthetic technique was individualized for each case due to complexity and rarity of the condition. The difficult airway risk factors were evaluated and an ENT might be called in cases of difficult ventilation and intubation. Results: A total of 20 patients aged 1.34 years old were anesthetized by the same anesthesiologist. In 10 cases it was decided to carry out sedation while in the other 10 cases there was need for general anesthesia. The airway management in cases of general anesthesia was performed with the use of laryngeal mask and tracheal intubation was not necessary. The inducing drugs used included ketamine and propofol, and anesthesia was maintained with sevoflurane. In cases of sedation we choosed the use of diazepam and dexmedetomidine. One patient had an episode of transient desaturation and one patient presented bradycardia which was reversed by atropine. No complications were observed in the other cases. Conclusions: To our knowledge this the largest case series of anaesthesia for MRI and diagnostic tests in MPS patients, which can be properly performed under sedation. In cases where general anesthesia becomes necessary, the laryngeal mask is a suitable alternative. The choice of anesthetic technique and the anesthesiologist.s experience in the management of these patients are critical to the success of the procedure and early recovery. Unitermos: Anesthesia; Mucopolysaccharidosis; Magnetic ressonance imaging