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## Anesthesia management in a patient with Freeman Sheldon syndrome

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**Introduction:** Freeman-Sheldon syndrome (FSS) or distal arthrogyria (whistling face syndrome, craniocarpotarsal dystrophy) is a rare congenital disease characterized by type 2 face and skeletal abnormalities. In patients with FSS, various anesthetic problems such as difficult airway, and difficulty of IV cannulation and regional interventions are encountered. In this report, anesthesia approach in case of FSS in whom operation is planned for circumcision and inguinal hernia repair is presented.

**Case:** A five year old boy at the weight of 12kg diagnosed with FSS underwent preoperative physical examination and mouth opening was restricted, head-neck extension comfortable and tiromental distance normal. In addition, in preoperative routine laboratory and imaging tests, all values were found to be within normal ranges. Prior to anesthesia, difficult airway and dantrolen sodium preparation was made. Anesthesia induction was made with 2mg/kg propofol and laryngeal mask was placed, controlling the airway without any problems. Anesthesia maintenance was made with TIVA. At the end of the operation, respiratory effort returned and anesthesia was terminated without any complications. For postoperative analgesia, 30mg/kg paracetamol was administered and the patient was transferred to recovery unit. The patient was transferred to his ward without any complications.

**Discussion:** Freeman Sheldon syndrome is a rare congenital disease belonging to distal arthrogyria group and characterized by dysmorphic structure combined with bone anomalies, joint contractures and typical facial appearance. In many cases, it arises sporadically without any family history. However, autosomal dominant and recessive cases have also been demonstrated. It occurs at equal rates in both sexes. Microstomy, distal arthrogyria in hand and feet accompanied by ulnar deviation, flexion contractures and resistant kinovarus are typical characteristics of the syndrome. Contracture in orbicularis oris muscle is the cause of 'whistling face' appearance. Although cases are present in the literature reporting muscular rigidity in FSS cases following halotan and succinylcholin administration, the relation between malignant hyperthermia and sevoflurane has not been definitively established so far. FSS cases undergoing successful anesthesia procedure with Sevoflurane without any finding of hyperthermia have been reported in the literature.

**Conclusion:** It is our suggestion that in FSS cases, one should be ready for difficult airway in anesthesia management, and the possibility of malignant hyperthermia should be borne in mind and LMA may be safely used in short lasting operations that do not require muscular relaxation.

### Biography

Ozkan Onal has completed his medical education at Gazi University Medical Faculty and he was specialized in anesthesiology in Hacettepe University Medical Faculty. He has more than 15 publications in reputed journals in the field of anesthesia.

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