CASE REPORT

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Anesthetic Management for Dental Procedures of a Child with Frank-ter Haar Syndrome

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ABSTRACT Frank-ter Haar syndrome is a very rarely encountered genetic disorder. Multiple skeletal abnormalities and abnormal facial features of these patients cause an increase in difficult intubation. This report presents our experience of a pediatric female patient with a difficult airway due to her Frank-ter Haar syndrome features. Patient repeatedly underwent two different dental procedures, under sedation and general anesthesia. The patient was prepped with soft cushions during operations. Intraoral secretions, bleeding and irrigation solutions were carefully suctioned during sedation. Another precaution taken was the disuse of muscle relaxant. Videolaryngoscope was another consideration used for the difficult intubation case. Both procedures were finalized without complications. Frank-ter Haar syndrome patients require wider range of planning and preparation in order to provide comfort and success.

Keywords: Frank-ter Haar syndrome; deep sedation; general anesthesia; airway management

Frank-ter Haar syndrome (FTHS) was initially described in 1973 by Frank et al. in three related children with severe cardiovascular complications and afterwards confirmed by ter Haar et al.[1-3] It is a rare autosomal-recessive genetic disorder, resulting from homozygous mutations in the SH3PXD2B gene on chromosome 5q35.1 coding for the Tks4 protein.[4,5]

FTHS is characterized by multiple skeletal, cardiovascular, and eye abnormalities, developmental delay and hypertelorism.[6,7] Brachycephaly, wide fontanels, prominent forehead ears and eyes, macrocornea, full cheeks, small chin, flexion deformity of the limbs, and prominent coccyx are important diagnostic features.[7]

It has been observed that in literature only 30 cases have been shared and amongst them reports investigating the dental features of these patients are scarce.[4,8,9] Köse et al. presented a 21-year-old male FTHS patient with gingival hypertrophy, broad alveolar crests, anterior open bite, micrognathia, bilateral condylar anomaly, a large right condylar process, elongated and widened styloid process, deep antegonial notches and multiple impacted teeth, predominantly the permanent upper and lower molars. The presented impacted teeth appeared to be in relation with dentigerous cysts.[4] Haznedaroglu et al. reported a 6-year-old male case with hyperplastic gingiva, and multiple caries.[8] Parker et al. reported follow-up of 3 FTHS patients.[9] Dental features of all three cases consisted of generalized gingival hypertrophy, delayed dental development, and multiple impacted teeth.

A single report could be reached, presenting the challenging anesthetic management of these patients.[10] Therefore, in the present case report, pa-

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tient and airway management during dental procedures of a FTHS case has been shared and discussed in light of present knowledge.

CASE REPORT

The local Institutional Review Board granted exemption in writing, because of the retrospective nature of the study. The study was conducted in accordance with the Declaration of Helsinki and parental and patient consents were obtained for publication of photographs.

A 6-year-old, (14 kg, 104 cm) female patient with FTHS was consulted for dental treatment regarding pain and swelling in the right mandibular molar region. The patient was previously diagnosed with FTSH. In medical history, the patient had additional mitral valve prolapse, moderate mitral insufficiency, mild aortic insufficiency and tricuspid prolapse. The patient was previously operated for patent ductus arteriosus closure. Physical examination revealed the classic features of FTHS consisting of growth retardation, severe kyphosis, narrow chest, shortened neck length, prominent forehead, ears and eyes, macrocornea, full cheeks, small chin, flexion deformity of the limbs. The patient had partial fixed neck deformity and neck movement was limited. Mouth opening was 2 cm and Mallampati score was 4 (Figure 1). SpO_2 was 97% in room air, chest auscultation was normal. Cardiac auscultation had systolic murmur (3/6). Echocardiography (ECG) showed sinus rhythm.

FIGURE 1: Physical examination of the patient.

In the intraoral examination, all permanent first molars were absent and pain on palpation and swelling was present in the right mandibular molar region. Radiographic evaluation revealed impacted first molars.

Sedation: Standard monitorization and pre-oxygenation was provided. The six-year-old patient was cooperative. Oral premedication was not used to be safer in respiratory danger. Anesthesia was induced with a mask by preserving spontaneous breathing with sevoflurane in 50% oxygen. Mask ventilation was easy. Peripheral vascular access and standard monitorization was provided. After administration of 2 mg×kg propofol, propofol infusion was continued. The depth of anesthesia was maintained so that the Ramsey sedation score was 5-6. The patient was oxygenized with nasal cannula. Because of the severe kyphosis and flexion deformity of the limbs, the patient was supported with pillows, from the waist, shoulders, knees and feet.

Local anesthesia (articaine HCl) was applied to the operation site. During the operation, infusion dose was regulated according to patient movements. A moist gauze was placed into the back of the tongue in order to prevent foreign bodies to enter the respiratory track and materials such as blood and irrigation solutions were suctioned carefully by an experienced technician. Before considering extraction of the impacted first molars, a more preservative approach was chosen and the mucosa layer over the permanent molar was removed under sedation in order to facilitate eruption. The procedure lasted for 25 minutes and was finalized without any respiratory or hemodynamic complications

General Anesthesia: The same case repeated 2 months later, and the patient was consulted again with pain and swelling in the same region with an additional extraoral fistula. Video-laryngoscope and fiberoptic bronchoscope was prepared for intubation. During the procedure, ECG, non-invasive blood pressure, SpO_2 and additional Bispectral Index (BIS) monitorization was provided. Because of the severe kyphosis and flexion deformity of the limbs, the pa-

tient was supported with pillows, from the waist, shoulders, knees and feet.

No premedication was given. Anesthesia was inducted with sevoflurane in 50% oxygen and 2 mcg remifentanil was added along with 2 mg×kg propofol. No muscle relaxant was used. While BIS was between 20-30, the patient was intubated in the first attempt (lasted 3 minutes) with endotracheal tube (No: 4, cuffed, reinforced ETT, Bıçakcılar, İstanbul, Turkey) using a video-laryngoscope (C-Mac, Karl Storz, Tuttlingen, Germany). Cormack Lahen score of the patient with macroglossia was 4. After the successful intubation, the operation was started and lasted for 45 minutes. The permanent first molar and the neighboring impacted second molar had to be extracted along with abscess drainage. Anesthesia was maintained with sevoflurane in 50% O_2-N_2O throughout the procedure. Sedation and general anesthesia attempt of the FTHS case was successfully obtained. The patient was extubated after the procedure and taken to the recovery room when spontaneous ventilation returned.

DISCUSSION

While the anesthesia management of even healthy patients can be difficult, the anesthesia management of patients with FTHS is a complete challenge. This challenging situation is formed because of the severe cardiovascular complications, full cheeks, small chin features of the patients. In the present case in addition to these features, severe kyphosis, narrow chest, shortened neck length was observed. Also, there was partial fixed neck deformity and neck movement was limited. Mouth opening was 2 cm and Mallampati score was 4. All of these situations alone are criteria that make airway management difficult, encountering all of them together makes the situation even more difficult.

The only report on airway management of a case with FTHS during dental procedures was by Tommasino and Albicini, for a 5-year-old male.[10] While the methodology of the anesthetic management was very similar, it should be emphasized that in the present case the patient was supported with soft pillows. The authors used this precaution due to contractures and posture disorders of the patient. Positioning with soft pillows will enable the medical staff to protect correct posture while increasing postoperative patient satisfaction. Patient support should be considered especially if a long-term procedure is planned. Suctioning of intraoral secretions, bleeding and irrigation solutions during sedation should also be highlighted.

Availability of technological products such as video laryngoscope and fiberoptic bronchoscope in such difficult intubation cases is crucial. In Tommasino and Albicini's case, they used the fiberoptic bronchoscope after an unsuccessful video laryngoscope attempt.[10] In the present case, success was achieved with video laryngoscope, nevertheless fiberoptic bronchoscope options were kept available.

In the presented case, premedication was not administered since the patient was very cooperative. The use of preoperative midazolam was preferred in Tommasino and Albicini's case. The authors argue that disuse of premedication is important for not extending the patient recovery time and that premedication should be only considered when patient cooperation is not present.

Depolarizing (succinylcholine) and non-depolarizing muscle relaxants are used for intubation in general anesthesia. Succinylcholine is used because of its rapid onset and very short duration of action features. Due to complications such as myalgia, increased intracranial pressure, hyperkalemia and malignant hyperthermia, today it has fallen behind in the preference list. Non-depolarizing muscle relaxant drugs are used as an alternative. However, they have slow onset, prolonged duration of action and prolonged reversing periods of their effects during failed intubation and airway management with mask ventilation and are associated with pulmonary complications of reversal agents.[11] The short-acting opioid remifentanil and propofol were chosen for their effectiveness in suppressing airway reflexes, and muscle relaxants were not used in intratracheal intubation. The same was applied in Tommasino and Albicini's case.

Although many precautions have been presented in the study, each case has its own characteristics, and the anesthetic managements should be considered case specific. For example, while general anesthesia is preferred for long-term dental interventions, seda-

tion should be preferred for shorter dental procedures. Mindful planning and thorough preparation are the key to provide patient comfort and a successful management for FTHS patients.

This report highlights the precautions taken during anesthetic management of a patient with FTHS. These precautions involve patient support with soft pillows, presence of video laryngoscope and fiberoptic bronchoscope options for intubation and the reasonable use of premedication and muscle relaxants.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

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