

CASE REPORT



Dental treatment in a hospital setting for a patient with Apert syndrome: A case report

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Abstract

Apert syndrome is a rare genetic syndrome resulting from missense mutations in the encoding fibroblast growth factor receptor 2. This case discusses the provision of routine dental care for a 22-year-old female with Apert syndrome. This patient presented with characteristic features of the syndrome including syndactyly, craniosynostosis, midface hypoplasia, and oral features such as dental crowding, impacted teeth, malocclusion, anterior open bite, and lateral swellings of the palatine processes. These characteristic oral and craniofacial findings provide unique challenges for dental practitioners. Proper management of this case required multidisciplinary care delivered in a hospital setting. Together, general dentists and oral surgeons performed extractions and routine prophylaxis without complication for this patient.

Keywords: Apert syndrome, Dental treatment, Lateral palatal swelling, Syndactyly

Introduction

Apert syndrome is a genetic syndrome caused by missense mutations in the gene that encodes fibroblast growth factor receptor 2 (FGFR2), leading to developmental abnormalities.^[1] The prevalence of this rare syndrome is reported to be one in 65,000 births.^[2] It is characterized by syndactyly, midface hypoplasia, and multisuture craniosynostosis (premature fusion of cranial sutures).^[1-3] Diagnosis relies on identification of the presence of these typical phenotypic features and can be further confirmed by molecular genetic testing to identify the pathogenic variant in FGFR2.^[2] While clinically similar to other syndromes with craniosynostosis, such as Crouzon, Pfeiffer, Muenke, and Saethre-Chotzen syndrome, patients with Apert present with the distinguishing characteristic of syndactyly in addition to a greater degree of midface hypoplasia than patients with Crouzon syndrome.^[3] Other clinical features include abnormalities of the skin, skeleton, and other organs. Brain abnormalities can be present, with mental effects ranging from none to severe.^[2]

Most pertinent to this case, patients who possess Apert syndrome have a variety of craniofacial abnormalities and distinctive oral characteristics.^[1-3] Proper dental management of these patients is imperative as they may face specific challenges to their oral health. Patients with Apert syndrome may have difficulty with dexterity or dental overcrowding.^[1] Surgical approaches

may be required during development to correct craniosynostosis, midface hypoplasia, and cleft palate.^[3] It is often necessary to deliver routine dental care during the patient's development in a hospital setting in conjunction with a team of other health providers, including but not limited to anesthesiologists, orthognathic surgeons, otorhinolaryngologists, orthopedic surgeons, orthodontists, plastic surgeons, and internists.^[1,3] This case report discusses an instance of delivery of routine dental care in a hospital setting.

Case Report

A 26-year-old female with diagnosed Apert syndrome and acute situational anxiety reported to the outpatient clinic for dental care. At the time of presentation, the patient exhibited severe dental crowding, malpositioned teeth, and gingival inflammation. Photos and a panoramic radiograph were taken at this time. The patient was unable to tolerate intraoral radiographs. Due to the patient's inability to tolerate dental treatment in a clinical setting, it was recommended her case be completed under general anesthesia at the outpatient surgical center.

The patient presented to the hospital with her parents, and pre-operative protocol was followed including history and physical review and obtaining a signed informed consent.

The patient was pre-medicated with midazolam 20 mg syrup by anesthesia and taken to the operating room where general anesthesia was induced through mask and a nasal endotracheal tube was placed. The head was wrapped, the eyes were lubricated and taped and the patient was draped in the usual manner. A throat pack was placed.

There were 17 intraoral radiographs taken. Comprehensive clinical examination and review of the radiographs revealed impacted and malpositioned teeth. Severe generalized gingival inflammation and a severely enlarged tongue were also noted.

The patient was treatment planned for the following: Tooth # M simple extraction, Tooth # R simple extraction, Tooth # 1 simple extraction, Tooth # 4 simple extraction, Tooth # 16 full bony extraction, Tooth # 17 full bony extraction, Tooth # 25 simple extraction, and Tooth # 32 full bony extraction.

Local anesthesia was achieved using 3.6 cc 2% lidocaine with 1:100,000 epinephrine and 6 cc 1% lidocaine with 1:100,000 epinephrine. The extractions of teeth #1, 16, 17, and 32 were completed by the oral surgery service. Teeth #M, R, and #4 were extracted by general dentistry. A dental prophylaxis and topical fluoride treatment were completed.

The oropharynx was suctioned and the throat pack was removed. The patient was then awakened, extubated, breathing on her own, and taken to the recovery room in good condition with an estimated blood loss of <5 cc. Her condition was stable and there were no complications. This patient tolerated the anesthesia and dental treatment well.

Discussion

Apert syndrome is a clinically distinct human condition characterized by craniosynostosis, craniofacial anomalies, and symmetric syndactyly of hands and feet, with cutaneous and bony fusions.^[4]

Common oral findings in individuals with Apert syndrome include dental anomalies, ectopic tooth positioning, supernumerary teeth, and tooth impaction.^[5] Children with this syndrome are often cared for by an interdisciplinary team to manage some of the dental anomalies associated with this condition. Dental anomalies may include tooth agenesis, typically the maxillary canines, and more than 40% of children with this syndrome also display enamel opacities. The maxillary first molars often erupt ectopically and patients often present with lateral palatal swellings. Abnormal occlusal relationships, delayed eruption, dental crowding, and missing teeth are some orthodontic challenges that require management.^[3] As evident in the panoramic radiograph in Figure 1, our patient presented with retained primary teeth, dental crowding, and impacted teeth [Figure 1].

Lateral swellings of the palatine process make the hard palate structure rare in this syndrome. The swellings are present in infants. As the child grows, the swellings increase in mass. These patients are sometimes misdiagnosed with cleft palate because a pseudocleft in the midline can develop from the cumulative

tissue proliferation.^[4] However, submucous and frank cleft palates have also been reported in patients with Apert syndrome, with one study showing an incidence of bifid uvula and cleft soft palate in 43.5% of patients with the specific S252W FGFR2 mutation.^[6] The typical palatal morphology of these patients is related to the maxillary hypoplasia observed in this syndrome. This hypoplasia leads to compression of the dental arch, which then becomes V shaped, giving rise to irregular tooth positioning and thickening of the alveolar ridge^[5] [Figure 2].

Almost all individuals with Apert syndrome display craniosynostosis. A majority of individuals with this condition are born with fusion of one or more cranial sutures. Overtime, other sutures may progressively fuse. Depending on which sutures are involved in this process, most children with this syndrome have a large anterior fontanelle that is displaced anteriorly onto the forehead. The most commonly involved sutures are the coronal (near 100%), sagittal (approximately 85%), and lambdoid (81%).^[3]

A diagnostic characteristic of individuals with Apert syndrome is fusion of the middle three digits of the hand. The fifth finger and thumb may be fused as well. Typically, a single nail (synonychia) is formed from the fusion of the fingernails for digits 2–4. Syndactyly of the toes may involve all digits, digits 2–5, or the lateral three digits. While both the upper and lower limbs can be affected, the upper limb is generally affected to a greater extent^[5] [Figure 3].

Individuals with Apert syndrome have a midface that is underdeveloped and retruded. A greater degree of vertical impaction leads to the maxillary bone being shorter in nature. Downslanting palpebral fissures and shallow orbits take form



Figure 1: Panoramic radiograph of the patient



Figure 2: Lateral palatal swellings (left photo taken 2015; right photo taken 2021)



Figure 3: Syndactyly



Figure 4: Clinical photograph of the face of the patient

by the underdevelopment of the midface. The appearance of relative mandibular prognathism and malocclusion is the result of maxillary structures not developing correctly^[3] [Figure 4].

The multidisciplinary team caring for a patient with Apert syndrome is ideally large and in constant communication. In certain circumstances, appointments and procedures can be combined to save the patient and family time and decrease the frequency of visits to the hospital or clinic.

Conclusion

Early intervention, awareness of the common craniofacial and dental presentations, and knowledge of their management are

important when formulating a treatment plan that best suits the patient's needs. The composition of the multidisciplinary team and excellent communication between these disciplines is imperative when caring for an individual with Apert syndrome.

Clinical Significance

This patient's oral health was most detrimentally impacted by malocclusion and dental crowding. Due to financial and behavioral constraints, the patient's caregivers elected not to undergo orthodontic care. Thus, exodontia of retained primary teeth, wisdom teeth, and selectively chosen permanent teeth was required to create space for improved oral hygiene. In an ideal scenario, orthodontic and surgical approaches may be used in conjunction to correct the defects seen in Apert syndrome.^[1] However, for this patient, exodontia with the aim of reducing dental crowding to improve oral hygiene and periodontal health was an appropriate and feasible treatment option.

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