Treatment Considerations for Failure of Permanent Teeth Eruption in Jacobsen Syndrome: A Case Report

Anna Ganios DMD, Gerald A Ferretti DDS MS MPH, Margaret E Ferretti DMD MPH, Ying An DDS PhD
Department of Pediatric Dentistry, University Hospitals Rainbow Babies and Children's Hospital
Case Western Reserve University School of Dental Medicine
Cleveland, OH

INTRODUCTION

Jacobsen syndrome is a rare genetic syndrome caused by partial deletion of chromosome 11. There is an estimated prevalence of 1/100,000 births with a female/male ratio 2:1. It is characterized by cognitive impairment, congenital heart defects and hematologic abnormalities. Characteristic facial features include skull deformities, hypertelorism, ptosis, downslanting palpebral fissures, small ears, broad nasal bridge, short nose and v-shaped mouth. Over two hundred cases of Jacobsen syndrome have been reported to date. This presentation discusses the management of a patient with Jacobsen syndrome in permanent dentition.

CASE REPORT

A 10 year old female presented to the University Hospitals Women and Children's Center Pediatric Dental Clinic for an appointment in December 2020. The patient's guardian was concerned that the permanent maxillary teeth had not erupted even though the primary teeth had exfoliated. The guardian reported the patient had periodic pain in the maxillary anterior area. Patient had no history of dental trauma and no family history of unerupted teeth.

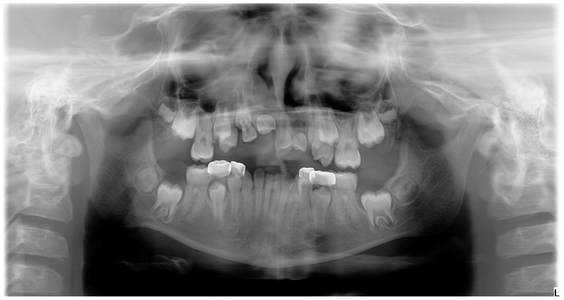
A clinical exam revealed that the patient was in mixed dentition, with presence of teeth 3, 13, 14, 19, K, L, 22, 23, 24, 25, 26, 27, S, T, and 30. The radiographic examination revealed no missing teeth, and teeth 4, 5, 6, 7, 8, 9, 10, 11, and 12 remained unerupted. The unerupted teeth had a clear path of eruption and were only covered by soft tissue. There was no clear etiology for the unerupted teeth.

The patient was referred to Case Western Reserve University School of Dental Medicine's Craniofacial Department in order to discuss treatment options for managing the unerupted teeth. The orthodontic team first placed a Hyrax expander to correct the transverse discrepancy. Furthermore, it was determined the patient requires a pacemaker. The medical team recommended all dental treatment be completed prior to pacemaker placement. Prior to the dental surgery appointment, teeth 4, 5, 11, 12, and 13 had erupted into the arch.

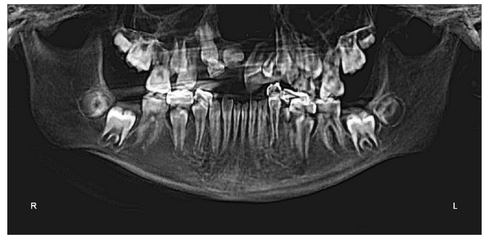
The patient was scheduled for comprehensive dental care under general anesthesia in November 2022 at University Hospitals Rainbow Babies and Children's Hospital. The patient required a platelet transfusion prior to the start of the surgery. The patient had sealants and composite restorations completed on teeth 3, 14, 19, and 24. The oral surgery team exposed teeth 6, 7, 8, 9, and 10. The patient was scheduled to take Amicar for five days post operatively.

CLINICAL AND RADIOGRAPHIC PRESENTATION

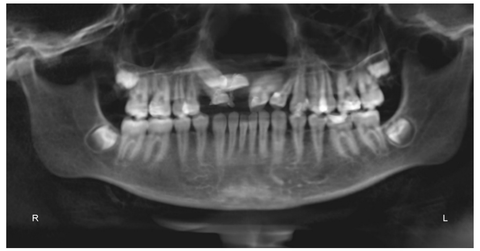
- **Initial presentation December 2020**



- **Presurgical presentation December 2021**



- **Postsurgical presentation January 2023**



DISCUSSION

Jacobsen syndrome is clinically characterized by dysmorphic features, delayed development of motor skills and speech, cognitive and behavioral impairment and congenital heart defects. There have been no specific dental manifestations reported in the literature for Jacobsen syndrome.

This case report presents a female with Jacobsen syndrome with unerupted permanent maxillary teeth due to unknown etiology. In addition, this case presents the challenges and a multidisciplinary treatment approach to facilitate care for the patient. There were concerns regarding function, esthetics, and psychological development due to failure of permanent teeth eruption. Thus, this dental presentation requires close monitoring with several specialists in order to obtain the best possible outcome. The long term plan is orthodontic treatment to improve alignment of the teeth once all permanent teeth erupted after surgical exposure.

The patient required comprehensive dental care under general anesthesia. Patient is currently followed by Case Western Reserve University School of Dental Medicine's Craniofacial Department for orthodontic treatment. Two months post surgery, teeth 7, 9 and 10 have erupted into the arch. Management of treatment expectations should also be considered as the remaining teeth may remain unerupted even with orthodontic treatment. Other treatment options including removable prosthesis, fixed appliances, or implant therapy will need to be considered.

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