Treatment Management of a Pediatric Patient with Complex Oligodontia

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BACKGROUND

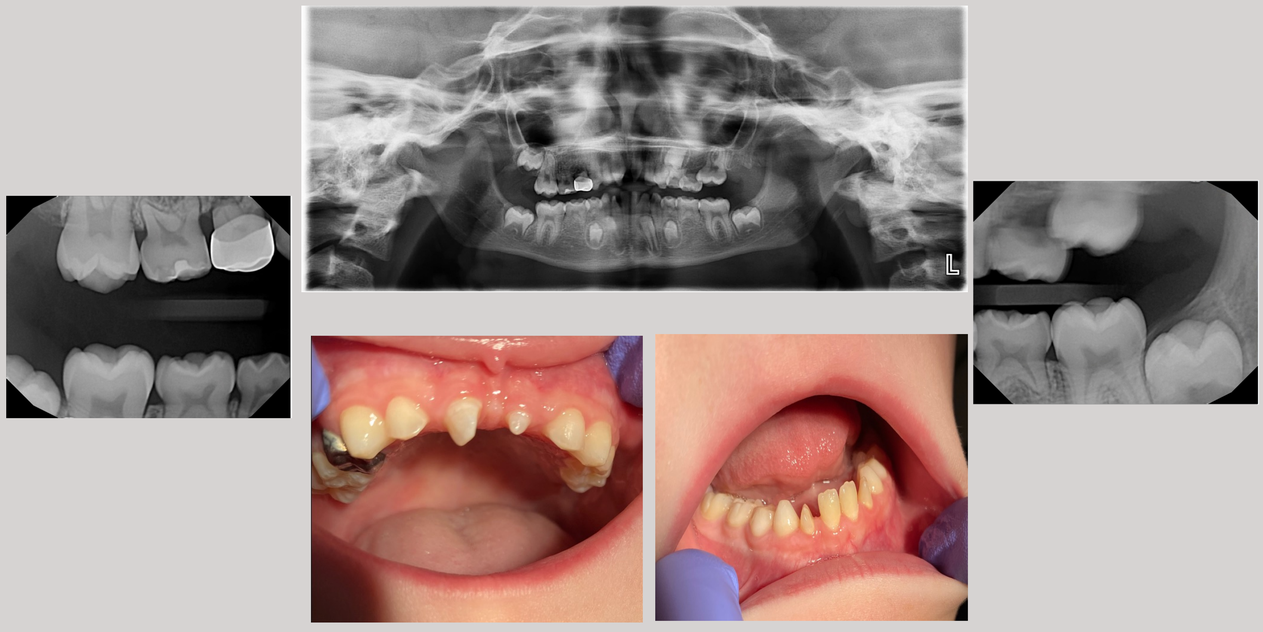
An 11-year, 3-month-old male presented to the Riley Hospital for Children outpatient dental clinic for comprehensive evaluation. His medical history was significant for Axenfeld-Rieger Syndrome (ARS), visual impairment, and pituitary gland deficiency for which he was taking Norditropin. He had no known drug allergies. ARS is a rare disorder inherited in an autosomal dominant fashion. It is estimated to occur in 1:200,000 persons and often goes undiagnosed for several years into childhood.[1] The pediatric dentist frequently plays a crucial role in the early diagnosis of ARS due to many of the morphological features involving dental and craniofacial anomalies. ARS originates as primarily an eye disorder with 50% of patients exhibiting glaucoma. This disorder is caused by a genetic mutation involving two known genes, PITX2 which results in type 1 ARS and FOXC1 resulting in type 3 ARS. The exact gene associated with type 2 ARS is not known but is believed to be located on chromosome 13.[1] A mutation involving the PITX2 gene frequently exhibits abnormalities of other parts of the body, not just the ocular region. A common dental presentation of patients with ARS consists of microdontia, oligodontia, enamel hypoplasia, short roots, and maxillary and mandibular hypoplasia.[1]

CLINICAL PRESENTATION

The patient's clinical examination indicated he was in mixed dentition, teeth #8 and #9 were microdonts, as well as there were other abnormally shaped and rotated teeth. The patient's mother was aware of the congenitally missing permanent teeth and was seeking opinions on how to best manage his complex presentation.

RADIOGRAPHIC INTERPRETATION AND DIFFERENTIAL DIAGNOSIS

Following interpretation of the patient's panoramic radiograph, it was determined that the patient is congenitally missing eight teeth (#4, 5, 7, 10, 13, 15, 20, and 29). He also has severe ectopic eruption of #14 that has completely resorbed the distal root of tooth #J, and a distal interproximal carious lesion on tooth #I involving the dentinal layer of the tooth.



MANAGING COMPLEX OLIGODONTIA

Oligodontia is defined as the agenesis of 6 or more teeth, excluding third molars.[2] Determining appropriate care for patients presenting with this can be complex, often requiring interdisciplinary care from various dental specialists including pediatric dentists, orthodontists, oral maxillofacial surgeons, prosthodontists, and periodontists.[3] Implant therapy is solely used in some cases, but typically bone levels are not adequate to successfully support implants. In this instance, as growth ceases pre-prosthetic preparation such as orthodontic therapy, orthognathic surgery, and osseous ridge augmentation are considered to achieve optimal function and esthetics for permanent prostheses.[3] Long term planning and early diagnosis of congenitally missing teeth, allows for early intervention with an interdisciplinary approach to establish a successful definitive treatment outcome, that allows the patient to achieve satisfactory appearance, oral function, and improved oral health related quality of life.[3]

DIAGNOSIS AND TREATMENT

A treatment plan was proposed to control all teeth exhibiting active disease and infection. This included extraction of tooth #B that had a periapical radiolucency as seen on the panoramic radiograph. Extraction of tooth #J was also warranted due to severe ectopic eruption of tooth #14.Following disease control, a consultation with an orthodontist is necessary to determine how to appropriately manage his congenitally missing teeth. After consultation with the Indiana University School of Dentistry orthodontic program, a decision will be made to address over retained primary teeth and how to most appropriately create and maintain space for future fixed prostheses. Following orthodontic extrusion of tooth #9 and correction of distally rotated central incisors, possible esthetic composite build ups of microdont #8 and #9 can be considered., until patient has reached his growth potential and can undergo bone augmentation and permanent prostheses.

LONG TERM MANAGEMENT OF ARS PATIENTS

Dental care should include regular clinical and radiographic monitoring and addressing chief complaints. Long term management by multidisciplinary approach, including speech and language therapy yields the best results.[4] Treatment of orofacial signs of ARS consists of correction of maxillary hypoplasia with myofunctional appliances or orthognathic surgery. Maintain primary teeth and correct crowding issues to maintain function and esthetics.[4] However, implant therapy remains the most likely long-term solution. It is imperative that pediatric dentists and oral surgeons are aware of ARS and its pathognomonic features, as the craniofacial abnormalities are early indicators of this syndrome, for better prognosis, quality of life, and prevention of various systemic complications.[4]

References:

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