Bilateral Supernumerary Primary and Permanent Maxillary Canines in Williams Syndrome

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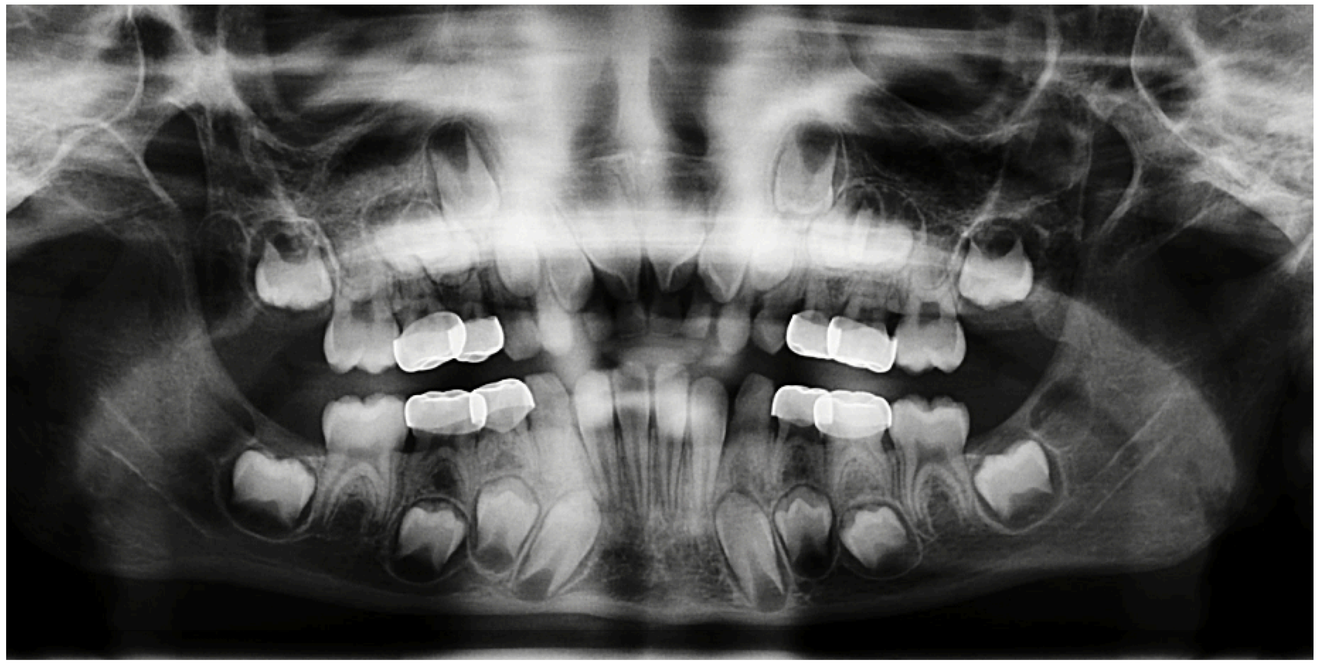
Introduction

Williams Syndrome (WS) is a rare genetic condition that is a result of the microdeletion on chromosome 7q11-23. Traditionally, patients with WS present with at least one dental developmental abnormality including diastemas, hypoplasia, and hypodontia.[2] In a study completed by T. Castro, et al. which evaluated oral characteristics in 52 subjects with WS, 51 were found to have some form of hypodontia.[1] In another study completed by Axelsson, Stefan, et al., 41 individuals with WS received dental evaluations and 40.5% experienced agenesis of at least one permanent tooth, while 11.9% experienced agenesis of 6 or more permanent teeth.[2] Due to these traditional dental anomalies, it is crucial that patients with Williams Syndrome receive early dental treatment.

Abstract

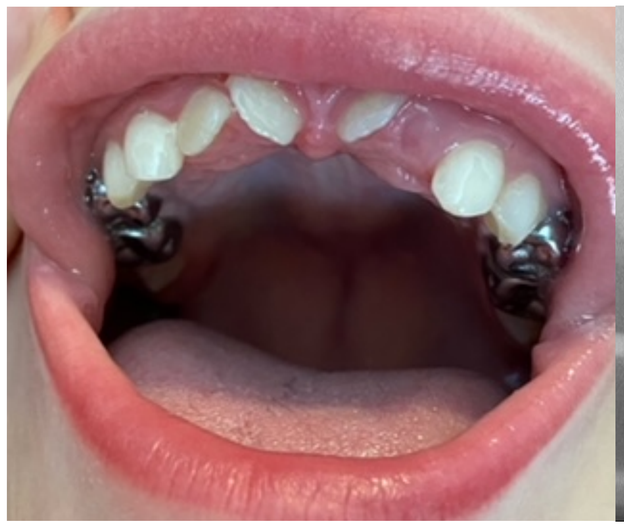
A 7-year-old male presented to Riley Children's Hospital dental clinic for routine recall examination. His health history included Williams Syndrome, aortic stenosis, hypertension, chronic otitis media, and development delay. The patient was taking 3 daily medications, Adderall XR, Enalapril, and Escitalopram. He had no known drug allergies. The patient received comprehensive dental treatment under general anesthesia in September 2018, including each primary molar receiving a stainless-steel crown. The patient presented in November 2021 for a recall examination when the patient was noted to have bilateral supernumerary primary and permanent maxillary canines. Considering patients with WS typically experience hypodontia rather than hyperdontia, this report includes radiographic and clinical findings documenting this unusual dental presentation in WS.

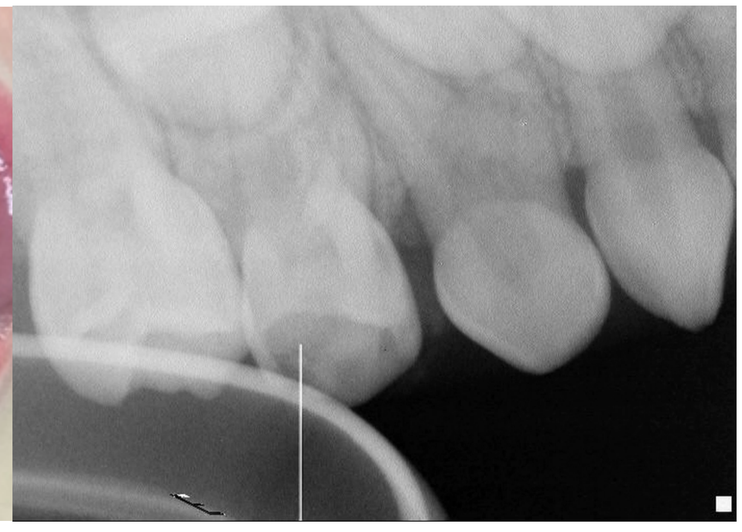


Radiographic Interpretation

Panoramic radiograph shows bilateral supernumerary maxillary primary canines and bilateral supernumerary maxillary permanent canines, representing four supernumerary canines.

Periapical radiograph shows a supernumerary maxillary right primary canine with two roots distal to #C.





Discussion

Due to the patient's rare dental anomaly, it is critical that the patient is monitored closely and frequently to observe dental development and ensure proper treatment in managing the developing occlusion. Due to the fact patients with WS often present with dental anomalies such as hypodontia, a patient with WS and hyperdontia is very rare, especially the presentation of four supernumerary canines. According to Anthonappa et al, the prevalence of a single supernumerary tooth is at most 3%.[3] Additionally, in a case report completed by Dos Santos et al claiming to be the first report of bilateral supernumerary primary and permanent maxillary canines, the authors state that the prevalence of a patient having one supernumerary canine is at most 1.5%. Dos Santos et al state that early diagnosis of supernumerary teeth is extremely crucial to minimize future complications.[4] This case report represents a patient with a medical history that would suggest the presence of hypodontia, when in fact the patient experiences the rare anomaly of four supernumerary teeth. Therefor, this case report illustrates the importance of proper clinical and radiographic exams in patients with syndromes.

References

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