

## INTRODUCTION

Coffin-Siris syndrome (CSS) is a rare genetic disorder characterized by developmental delay, craniofacial abnormalities, and multiple congenital anomalies associated with mutations in the BAF-complex or SOX pathway. First described in 1970, CSS presents with variable features including digital anomalies, intellectual disability, and systemic involvement. Due to its rarity and clinical variability, oral and dental manifestations are not well defined.

Tooth development and eruption rely on coordinated genetic and developmental processes throughout childhood and adolescence. Disruptions in these pathways may lead to altered dental maturation and eruption, contributing to functional, esthetic, and treatment challenges.

This case report describes the oral findings and dental management of an adolescent with Coffin-Siris syndrome presenting with delayed dental development, multiple over-retained primary teeth, and severe attrition. It highlights the importance of early evaluation, interdisciplinary care, and long-term follow-up.

## CASE REPORT

A 17-year-old male presented to University Hospitals Rainbow Babies and Children's Hospital for comprehensive dental rehabilitation under general anesthesia in March 2025. The patient was initially evaluated in October 2024 following referral from an outside dentist, with the chief complaint that multiple primary teeth required extraction due to delayed exfoliation.

The patient's medical history was significant for Coffin-Siris syndrome, agenesis of the corpus callosum, heart murmur, epilepsy, and asthma. Behavioral limitations and medical complexity contributed to reduced cooperation, preventing acquisition of panoramic imaging and a complete occlusion assessment in the clinical setting.

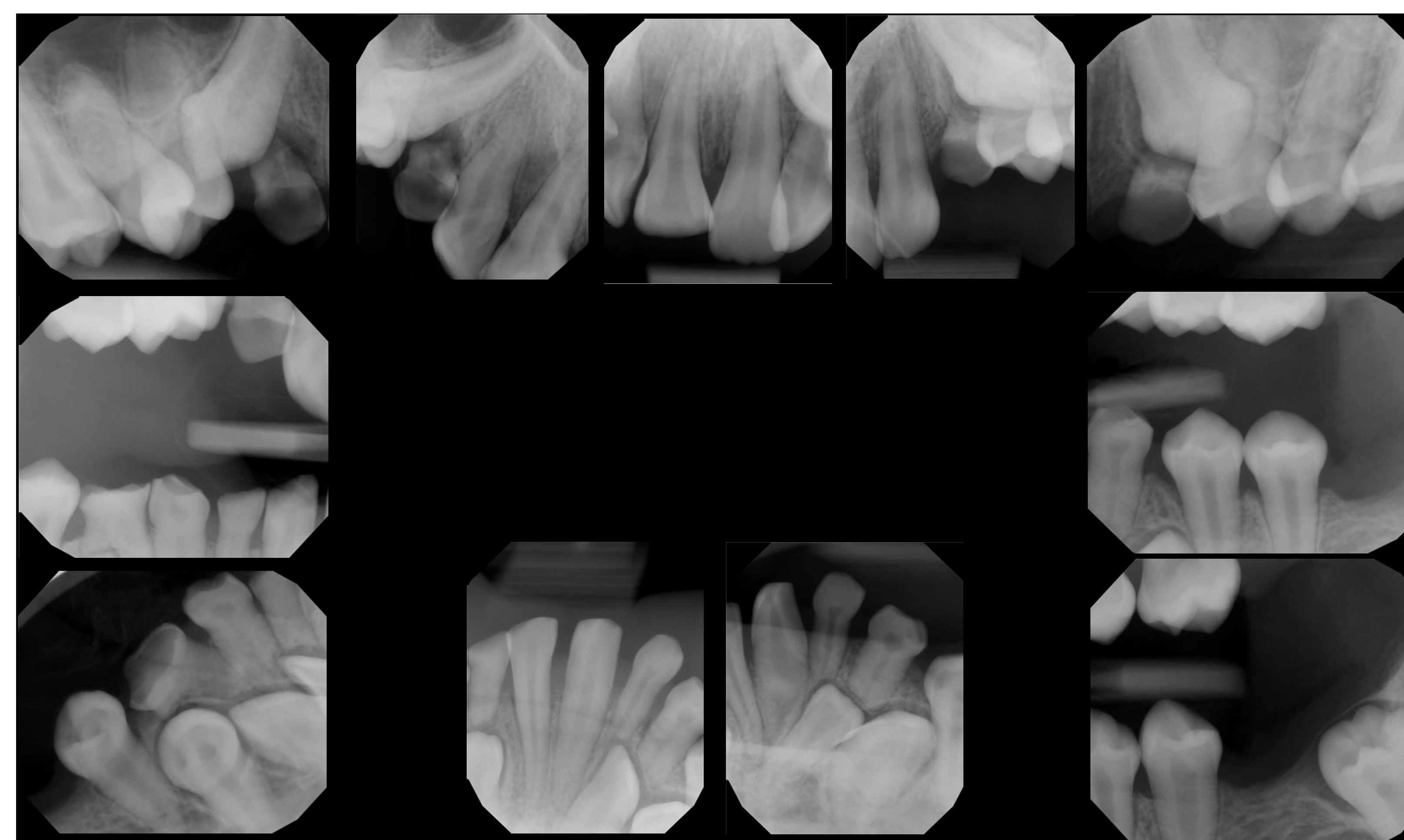
Intraoral examination revealed three carious permanent teeth, seven over-retained primary teeth, and generalized gingivitis associated with plaque accumulation. Retained primary teeth (#C, H, M, N, Q, R, S) exhibited severe attrition approaching the pulp and were notably non-mobile, indicating lack of normal exfoliation. The patient's dental age was significantly delayed relative to chronological age, supported by the reported eruption of maxillary anterior teeth at approximately 10–11 years of age. Clinical findings raised concern for altered eruption patterns and possible absence of permanent successors, including teeth #18 and #31; however, radiographic confirmation was not obtained.

Given the extent of attrition and poor prognosis of select retained primary teeth, teeth #C, H, M, N, and Q were extracted. Additionally, composite restorations were completed on teeth #3-O, #14-O, and #29-OB to address carious lesions and maintain function.

## CLINICAL PRESENTATION



## RADIOGRAPHIC PRESENTATION



## DISCUSSION AND CONCLUSION

Providing dental care for individuals with Coffin-Siris syndrome can be complex due to the combination of systemic conditions, developmental delay, and challenges with patient cooperation. This case demonstrates several notable dental findings, including significant delay in tooth eruption, persistence of multiple primary teeth, and severe attrition affecting the remaining dentition.

Normal dental development relies on coordinated genetic signaling and growth over time. In patients with syndromic conditions such as Coffin-Siris syndrome, these processes may be altered, resulting in deviations from typical eruption patterns and tooth development. The delayed dental age observed in this patient, along with the history of late eruption of the maxillary anterior teeth, aligns with previously reported findings of delayed eruption and other developmental dental anomalies in this population.

By late adolescence, eruption of the permanent dentition is generally complete, aside from third molars. In this case, the continued presence of multiple non-mobile primary teeth suggests disruption in the normal exfoliation and eruption sequence, raising concern for compromised or absent development of the permanent successors. These findings indicate a potentially limited prognosis for normal eruption of the remaining permanent teeth.

Management considerations for patients with significant delays in dental development must be individualized and consider both current function and long-term outcomes. Retaining primary teeth may be beneficial in maintaining alveolar bone and occlusal stability, particularly when permanent tooth development is uncertain. At the same time, the presence of severe attrition and caries risk complicates long-term planning and requires ongoing monitoring.

Given the rarity of Coffin-Siris syndrome, there is a lack of comprehensive data regarding dental presentation and long-term management. This case contributes to the limited body of literature and reinforces the importance of early diagnosis, regular dental follow-up, and a multidisciplinary approach to care.

## REFERENCES

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