# Oral Rehabilitation of a Patient With William's Syndrome: A Case Report

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## INTRODUCTION

Williams Syndrome (WS) is a chromosomal disorder caused by a deletion on chromosome 7q11.23 associated with the elastin gene. WS is usually identified by diagnosing the cause of specific cardiac abnormalities. Those with WS frequently have the following characteristics: cardiac malformations, kidney problems, increased risk of diabetes, cognitive and developmental abnormalities, and connective tissue irregularities.

Common physical characteristics can include facial dysmorphisms or elfin facies, and dental malformations such as malocclusion, missing and malformed teeth, pulp stones, increased space between teeth, enamel hypoplasia, and high prevalence of dental caries

## CASE REPORT

A 15-year-old male with the primary diagnosis of Williams syndrome (WS) presented to the University Hospitals Rainbow Babies and Children Hospitals dental clinic in June 2024 as a referral due to dental treatment requiring sedation with cardiac anesthesia. The patient's medical presentations included: pulmonic valve stenosis that had been resolved, hypercalcemia, vision deficits, short stature, and elfin-like features (facial dysmorphism). Genetic testing confirmed the diagnosis of WS after the patient had cardiac complications, and the known diagnosis of WS in the patient's father.

Craniofacially, the patient does not present with the typical syndromic features as seen in many with WS. He has a convex facial profile with long lower anterior facial height and strained mentalis. He is symmetrical bilaterally and presents with a broadened mouth and full lips, as well as lip incompetence related to malocclusion (see figure 1 A-B). Intraoral exam showed the patient was in mixed dentition, with excess interproximal spacing, and an anterior open bite. He is congenitally missing teeth #1,4,13,16,20,29 (see Figure 2 A). Teeth # A,J,T were over retained, carious, and presumed to be ankylosed (Figure 2A-E). The patient's teeth # 7 and 10 appeared to be peg laterals (Figure 2A). In addition, #3,14,30 were grossly decayed and non-restorable. On clinical and radiographic exam, restorable decay was also noted on #5,8,9,12,15,18,19,28, and 31. Due to the patient's health history and inability to sit for procedures, all treatment was completed at UH Rainbow Babies and Children's Hospital in October 2024. The dentition was restored with composite. Pediatrics partnered with Oral Surgery following the completion of the necessary restorations to remove impacted, ankylosed, and grossly decayed teeth.

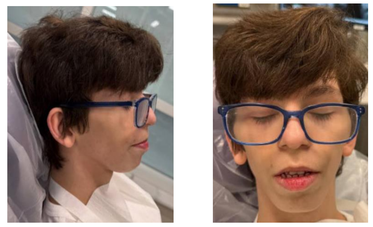

**Figure 1:** Extraoral photos taken during initial clinical consultation. Notice the evident appearance of the broadened mouth and full lips, as well as the convex profile, strained mentalis and, the anterior open bite.

## CLINICAL AND RADIOGRAPHIC PRESENTATION

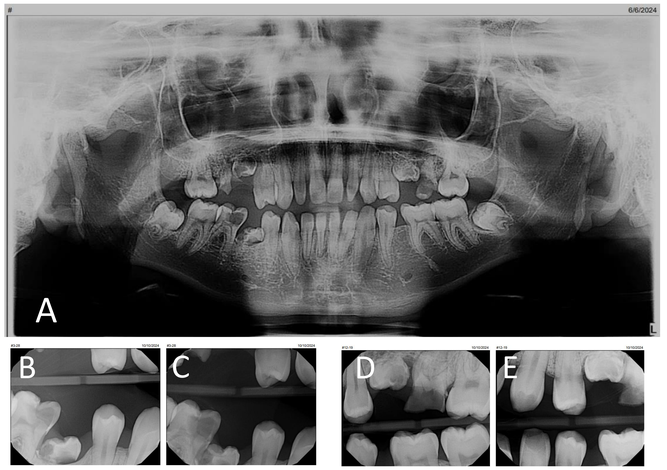


**Figure 2:** Pre-op panoramic (taken June 6, 2024) and bitewing radiographs (taken under anesthesia Oct. 10, 2024)

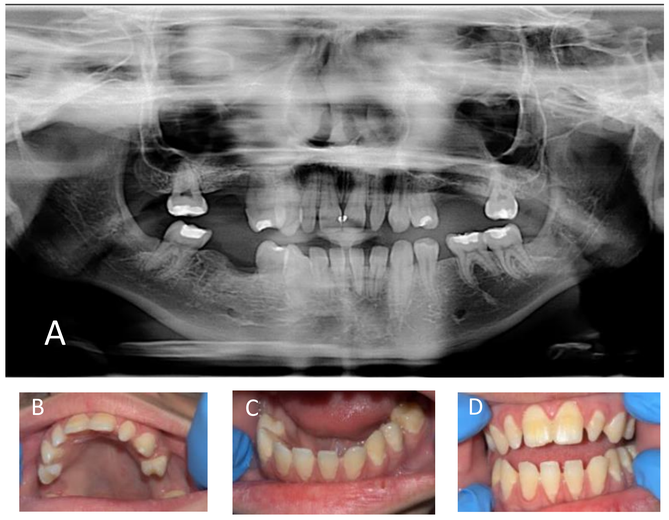


**Figure 3:** Post-op panoramic radiograph and intraoral photos were taken Dec. 30, 2024

## DISCUSSION

A 3-month follow-up showed appropriate healing of surgical sites (see Figure 3A-D) in December 2024. The gingiva is erythematous with slight gingivitis from plaque accumulation. The open bite malocclusion and excess spacing is evident. No specific areas of discomfort were reported. The patient and caregiver will be encouraged to improve hygiene which will be followed by a referral to the craniofacial orthodontics department at Case Western Dental School.

The case underscores the necessity of a collaborative healthcare approach, integrating pediatric cardiology, dentistry, and anesthesia teams. Cardiovascular assessment is crucial due to the high prevalence of supravalvular aortic stenosis and pulmonary stenosis in WS patients, which increases the risk of surgical complications. Routine dental monitoring is essential to manage the increased susceptibility to dental anomalies, including malocclusions, enamel hypoplasia, and high caries risk. Specialized anesthetic considerations are necessary due to the heightened risk of perioperative cardiovascular instability. Individuals with WS may also exhibit neurological and behavioral traits, including hypersociability, anxiety disorders, and attention deficits, which require tailored management approaches. Structural brain abnormalities, particularly in the dorsal-frontal pathways, contribute to the unique neurocognitive profile of WS patients. Gastrointestinal manifestations, including chronic constipation and increased risk of diverticulitis, are also prevalent and should be managed accordingly. Connective tissue abnormalities, such as joint laxity and spinal deformities, necessitate orthopedic evaluations, and musculoskeletal monitoring.

Williams Syndrome presents a complex interplay of medical and dental challenges, requiring specialized care coordination. The case highlights the importance of early diagnosis, individualized treatment plans, and interdisciplinary collaboration to optimize patient outcomes. Comprehensive management strategies should address the diverse medical, developmental, and psychosocial needs of affected individuals. Future research should focus on long-term cardiovascular and dental management strategies to enhance the quality of life for individuals with WS.

## REFERENCES


- American Heart Association. (2013). Hypertension in patients with Williams Syndrome. *Hypertension, 62*(1), 7-16
- Morris CA. Williams Syndrome. 1999 Apr 9 [updated 2023 Apr 13]. In: Adam MP, Feldman J, Mirzaa GM, Pagon RA, Wallace SE, Amemiya A. University of Washington, Seattle; 1993–2025. PMID: 20301427
- Pober, B. R. (2010). Williams-Beuren syndrome. *American Journal of Medical Genetics Part C: Seminars in Medical Genetics, 154C*(2), 203-215.
- Wilson M, Carter IB. Williams Syndrome. [Updated 2023 Jun 26]. Treasure Island (FL): StatPearls Publishing; 2025 Jan.
- Wong D, Ramachandra SS, Singh AK. Dental management of patient with Williams Syndrome - A case report. Contemp Clin Dent. 2015 Jul-Sep;6(3):418-20. doi: 10.4103/0976-237X.161908. PMID: 26321847; PMCID: PMC4549999.