



Introduction

Sotos syndrome is a rare genetic overgrowth disorder characterized by distinctive craniofacial features, developmental delay, and excessive physical growth during childhood. The condition is most commonly caused by pathogenic variants in the NSD1 gene and is typically inherited in an autosomal dominant pattern, although most cases arise from de novo mutations⁽¹⁾⁽²⁾. In addition to overgrowth and intellectual impairment, patients may present with systemic manifestations such as cardiac anomalies, renal abnormalities, and seizures⁽¹⁾⁽³⁾. Craniofacial and oral findings including high-arched palate, premature eruption of teeth, and dental anomalies have also been reported⁽⁴⁾⁽⁵⁾. These characteristics, combined with developmental delay, may increase the risk of dental disease and complicate dental management. This report describes comprehensive dental rehabilitation under general anesthesia in a child with Sotos syndrome.

Case Report

A three-year-ten-month-old female diagnosed with Sotos syndrome associated with an NSD1 gene mutation presented to the pediatric dental clinic for evaluation of multiple dental caries. Her medical history included developmental delay and febrile convulsions, and a family history of epilepsy was noted in a cousin. Growth records demonstrated accelerated growth consistent with Sotos syndrome; at two years and three months of age, her height was 96.4 cm, corresponding to approximately the 97th percentile for her age. At the time of dental consultation, her body weight was 21 kg.

Intraoral examination revealed multiple carious lesions involving several primary teeth, some with suspected pulpal involvement. A dental anomaly was also observed, consisting of fusion of the mandibular right primary central and lateral incisors. Because of the patient's young age and limited cooperation related to developmental delay, comprehensive dental treatment under conventional chairside conditions was considered difficult.

After discussion with the patient's caregivers, full-mouth dental rehabilitation under general anesthesia was planned in order to complete treatment safely in a single session. The patient subsequently underwent comprehensive dental rehabilitation, including pulpectomy for primary teeth with pulpal involvement and restoration of severely affected teeth using stainless steel crowns and zirconia crowns. The fused mandibular incisors were incorporated into the overall restorative treatment plan. The procedures were completed successfully without complications. Following treatment, the patient was scheduled for regular recall visits with preventive care including oral hygiene instruction, dietary counseling, and topical fluoride application.

Clinical findings



Figure 1. Initial intraoral examination



Figure 2. Finish intraoral examination

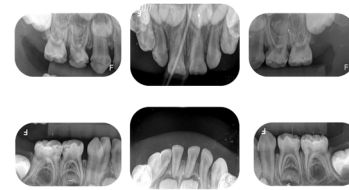


Figure 3. Initial radiographic examination

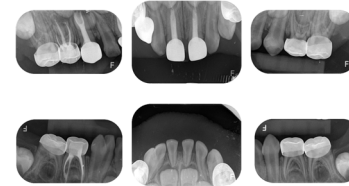


Figure 4. Finish radiographic examination



Figure 5. Ten months follow up radiographic examination

Discussion

Patients with Sotos syndrome often present with craniofacial and oral manifestations that may influence dental development and oral health. Developmental delay and behavioral difficulties frequently associated with the syndrome may further compromise oral hygiene practices and increase susceptibility to dental caries. In the present case, the patient exhibited severe early childhood caries affecting multiple primary teeth with pulpal involvement, requiring comprehensive dental management. The presence of a fused mandibular primary incisor also represented a dental anomaly that required consideration in treatment planning.

Dental treatment for young children with developmental delay can be challenging because limited cooperation may restrict the feasibility of conventional chairside procedures. When extensive dental treatment is required, comprehensive dental rehabilitation under general anesthesia provides a safe and efficient approach that allows multiple procedures to be completed in a single session. In this case, pulpectomy and full-coverage restorations with stainless steel and zirconia crowns were successfully performed under general anesthesia. Following treatment, regular recall visits and preventive strategies—including oral hygiene instruction, dietary counseling, and topical fluoride application—were emphasized to reduce the risk of recurrent caries and to maintain long-term oral health.

Conclusion

Children with Sotos syndrome may present with significant dental treatment needs due to craniofacial anomalies, developmental delay, and increased risk of dental caries. Comprehensive dental rehabilitation under general anesthesia can provide a safe and effective approach for managing extensive dental disease when patient cooperation is limited. Early diagnosis, preventive strategies, and regular follow-up are essential to maintain long-term oral health in these patients.

References

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