# Dental Rehabilitation of Pediatric Patient with Ellis-van Creveld Syndrome: Case Report

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## ABSTRACT/INTRODUCTION

Ellis-van Creveld Syndrome, also known as Chondroectodermal dysplasia, is a rare autosomal recessive disorder caused by mutations in the EVC and EVC2 genes[4]. This syndrome is characterized by polydactyly of the hands, short limbs, ectodermal dysplasia, and congenital heart defects. Because Ellis–van Creveld syndrome affects all three embryonic layers, it presents with a range of oral manifestations, including absence of the mucobuccal fold, a serrated alveolar ridge, multiple labial frenula, abnormalities in tooth morphology, and congenitally missing teeth[1]. This report details a 20-month-old female who presented to the Children's Mercy Dental Clinic in May 2019 for comprehensive dental care. The purpose of this report is to review dental treatment considerations for patients with Ellis-van Creveld Syndrome.

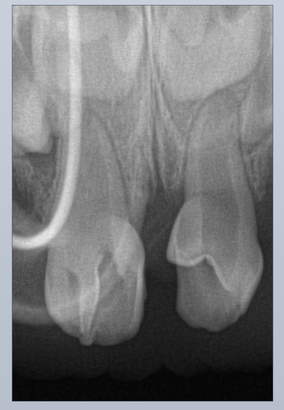

Figure 1

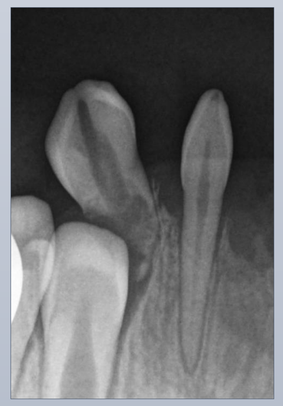

Figure 2

## CASE REPORT

A 20-month-old female initially presented to the Children's Mercy Dental Clinic in May 2019 to establish care. Medical history included Ellis-van Creveld Syndrome, developmental delay, ulnar polydactyly of fingers, patent foramen ovale with atrial enlargement, and eczema. Exam findings included decay on the first primary molars, an irregular appearance of the primary central incisors, and a broad maxillary frenum. Due to patient age, it was decided to place SDF on the carious first molars and wait until the primary second molars erupted before completing dental rehabilitation in same day surgery (SDS).

In November 2019, a periodic exam revealed several teeth with malformed enamel and non-typical morphology including geminated #R and #F which had a talon cusp and lingual pulpal exposure (Figures 1-2). Other exam findings included occlusal decay on #B, I, L, S, the lack of a maxillary vestibule, and mild gingivitis. At that time, SDS was recommended and completed in April 2020. Treatment included stainless steel crowns (SSCs) on primary posterior teeth and composite resins on #E and #F.

In January 2023, the patient presented for a routine dental exam. Findings included a missing crown on #A which had recurrent decay (Figure 3), congenitally missing #23, 24 26, geminated #25, multiple frenulum attachments, and moderate calculus build up. SDF was placed on #A due to inadequate space to recement the crown in clinic. Due to limited tolerance for in-office care and medical complexity, SDS was recommended again.

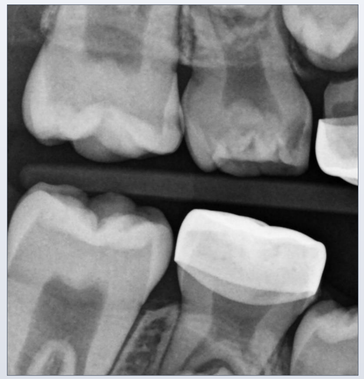

Figure 3

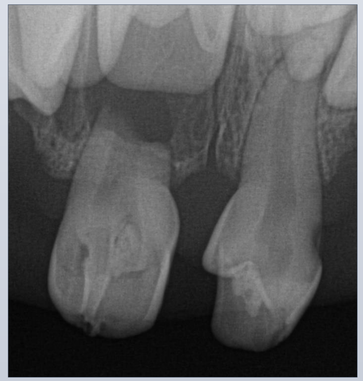

Figure 4

In April 2024, the patient presented for an emergency visit. Mom reported that the patient's top front tooth was loose and causing significant pain. Clinical exam showed a periapical radiolucency on tooth #E which was also class II mobile (Figure 4). #E also presented with dens evaginatus. A possible supernumerary tooth was noted near the developing crown of #9. Due to pulpal necrosis and symptoms, it was determined that tooth #E should be removed. Extraction was completed in clinic and SDS orders were resent.

In October 2025, dental rehabilitation in SDS was completed, including SSCs on #A, 3,14; an extraction of #B, C, F, I, L, M, R; and sealants on #19, 30. A delay in scheduling was caused by difficulties contacting the patient's guardian. The patient has not yet returned for follow up.

## DISCUSSION/CONCLUSION

Ellis-van Creveld syndrome is a rare genetic disorder that significantly disrupts normal dental development. There are several unique dental manifestations of Ellis-van Creveld syndrome, including an absent or shallow mucobuccal fold, multiple broad labial frenula, and teeth with abnormal morphology[1]. These anomalies, particularly hypoplastic and malformed teeth, can place patients at a higher risk for dental decay[5]. In this case, the patient presented with several of these characteristics which, in addition to her complex medical history, contributed to the need for comprehensive dental care in SDS.

Dental professionals must recognize the oral manifestations of Ellis-van Creveld syndrome and provide timely preventative and definitive care. Preventative care should include pit and fissure sealants, routine prophylaxis, and topical fluoride application. Definitive care may include SSCs, extractions, or composite restorations. In some cases, a partial or fixed denture may be indicated to improve speech, esthetics, and mastication[2,3]. Surgical correction may also be indicated for soft tissue and skeletal abnormalities[2].

This case highlights how gaps in dental care can result in an increased need for hospital dentistry, more aggressive intervention, and a greater risk for pain and infection. Routine dental exams are critical for monitoring development and guiding parents on oral hygiene and anticipatory care.

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