



## ABSTRACT/INTRODUCTION

Dandy Walker Syndrome is a rare congenital brain malformation in which the cerebellum does not fully develop. The cerebellum is responsible for controlling movement and balance and is located at the back of the brain<sup>1</sup>. Clinical symptoms associated with this syndrome include developmental delays, hydrocephalus, eye abnormalities and dental abnormalities including malformed or missing teeth<sup>1</sup>. Orofacial Digital Syndrome is another rare genetic disorder characterized by a combination of abnormalities affecting the face, mouth, hands and feet<sup>2</sup>. Clinical features of this syndrome include cleft lip, cleft palate, syndactyly, polydactyly, hypodontia and supernumerary teeth<sup>2</sup>.

This presentation discusses a 9-year-old female patient who has been seen at Children's Mercy Hospital due to her complex medical history. Her medical history is notable for Dandy Walker Syndrome, Orofacial Digital Syndrome Type 1, developmental delays and hydrocephalus. The patient does not have a specific chief complaint. Her mother is aware of her conditions and wishes to establish a dental home at a hospital equipped to provide necessary care. During a periodic exam, radiographic findings revealed the presence of supernumerary teeth and congenitally missing teeth. This report will detail the dental findings and treatment management for patients with Dandy Walker Syndrome and Orofacial Digital Syndrome.

## CASE REPORT

A 9-year-old patient initially presented to the Children's Mercy Dental Clinic in 2018 to establish a dental home. Patient previously had not been seen by a dentist. Medical history includes Dandy Walker Syndrome, Orofacial Digital Syndrome, developmental delays and hydrocephalus. At patient's initial exam her teeth were noted to be maligned and varied in shape and size. She did not have all her primary teeth and some of the primary teeth presented with a conical shape. No caries were detected at the initial examination and no treatment was recommended.

In September of 2020, patient presented for a recall exam and more dental anomalies and decay were noted. Radiographs were obtained and revealed three supernumerary teeth (#CS, #HS, TS), fused #O/P, and missing #N (Figure 1). Decay was noted on a few of the primary anterior teeth. SDF was offered as a treatment option, but Mom declined due to the dark staining it would cause. It was decided that treatment would be attempted in the clinic using nitrous oxide. Provider was successfully able to treat one cavity, but due to patient cooperation, no other treatment was able to safely be completed. Patient is not a Children's Mercy sedation candidate due to her ASA Class III classification. It was decided that treatment under general anesthesia would be the best option to ensure successful completion of treatment.

In February of 2024, patient received full mouth dental rehabilitation completed under general anesthesia. There were delays in getting patient scheduled for treatment including caregiver initially declining treatment with full coverage stainless steel crowns and then accepting the treatment plan due to patient's increased need for care and increased pain due to the extent of decay. Nine primary teeth were extracted including three supernumerary teeth (Figure 2). One permanent tooth received a composite restoration.

In October of 2025, patient was seen for a recall examination. Radiographs were obtained including a panoramic radiograph (Figure 3). Radiographs revealed more dental anomalies: supernumerary teeth and congenitally missing teeth. Patient is missing #23, 26 and has two supernumerary premolars #54, 79. Patient was found to have more decay, and general anesthesia was recommended for treatment due to her high dental anxiety and medical history. The patient has not been seen in same day surgery for treatment.

Figure 1: September 9, 2020



Figure 2: February 2, 2024



Figure 3: October 6, 2025



## DISCUSSION/CONCLUSION

Dandy Walker Syndrome is a rare congenital brain malformation characterized by an enlargement of the posterior fossa, dilation of the fourth ventricle and partial absence or hypoplasia of the cerebellar vermis<sup>3</sup>. Dental manifestations include malformed or missing teeth, as seen in this patient. Patients are typically diagnosed with this syndrome within their first year of life. Patients present with signs and symptoms of increased intracranial pressure, related to hydrocephalus<sup>3</sup>. Treatment for the intracranial pressure includes surgery where shunts are placed in the brain to help decrease pressure<sup>3</sup>. Dental treatment often includes full coverage restorations to help preserve malformed teeth which are more prone to decay.

Orofacial Digital Syndrome is characterized by oral, facial and digital anomalies usually diagnosed at birth. The Syndrome mostly affects females and is usually lethal to males during gestation<sup>4</sup>. Dental manifestations include a cleft palate, missing teeth, extra teeth, enamel dysplasia and malocclusion<sup>4</sup>. This patient did not have cleft palate, but she had missing teeth, extra teeth, enamel dysplasia and malocclusion. Treatment for these patients includes surgical repair for the cleft palate, orthodontics for malocclusion and full coverage restorations to help preserve malformed teeth.

Dental professionals should recognize the oral manifestations of Dandy Walker Syndrome and Orofacial Digital Syndrome and provide preventive care for these patients. This case highlights the treatment planning and management challenges that can arise when a patient presents with both syndromes and associated dental anomalies. Delays in care can result in the need for more extensive treatment and the need to complete the treatment in a hospital-based setting under general anesthesia. Routine dental care is essential in these patients, and timely, definitive treatment plays a critical role in promoting and maintaining positive oral health outcomes.

## REFERENCES

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