Female with Dandy-Walker Syndrome and Cleft Lip and Palate: A Case Report

Tonya Ozbeki, DDS , Rodney Vergotine DDS

University of Michigan School of Dentistry, Department of Orthodontics and Pediatric Dentistry

INTRODUCTION

Dandy-Walker syndrome (DWS) is a congenital brain malformation primarily affecting the cerebellum. First described by Dandy and Blackfan, and later expanded upon by Taggart and Walker, an estimated incidence of 1 in 25,000 to 35,000 live births in the United States.

The hallmark features of DWS:

- Enlargement of the fourth ventricle
- Partial or complete absence (agenesis) of the cerebellar vermis
- Formation of a cyst near the fourth ventricle
- Hydrocephalus leading to progressive enlargement of the skull

Neurological symptoms can include:

- Delayed motor development
- Cerebellar dysfunction resulting in ataxia
- Increased intracranial pressure manifesting as irritability, vomiting, and behavioral changes

DWS is frequently associated with other brain malformations such as agenesis of the corpus callosum. Systemic anomalies, including cleft palate, were present in approximately 25% of cases. DWS impacts multiple domains of development, particularly motor skills and behavior, necessitating a multidisciplinary approach to diagnosis and management

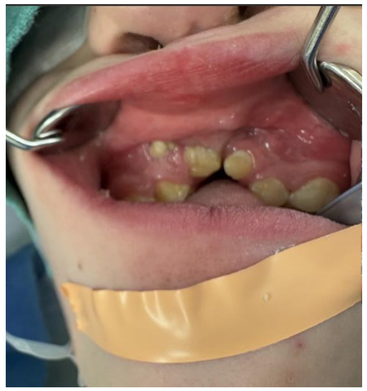
PATIENT INTRODUCTION

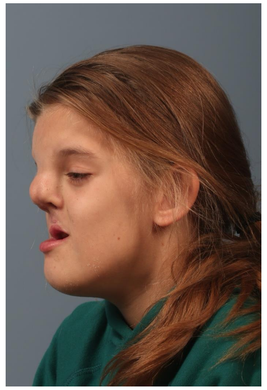
A 15y 7m female patient presented in November 2024 reporting that she has been experiencing dental pain. The patient is receiving ongoing care at C.S. Mott Children's Hospital-Dentistry Clinic as well as the Craniofacial Anomalies Team Clinic.

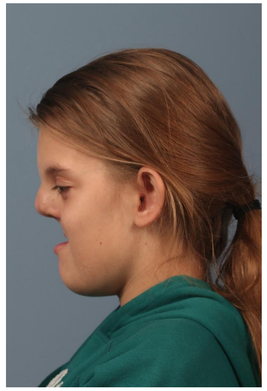
Past Medical History: Dandy Walker syndrome, Agenesis of corpus callosum, Frontonasal dysplasia, Cerebral palsy, Autism, Developmental delay, Bilateral cleft lip/palate s/p repair, ETD s/p tubes, Developmental articulation disorder, Hypertelorism, Mixed receptive-expressive language disorder, Hyperopia of both eyes with astigmatism, recurrent epistaxis, speech disturbance, PDA and PFO which spontaneously closed, history of infantile seizures

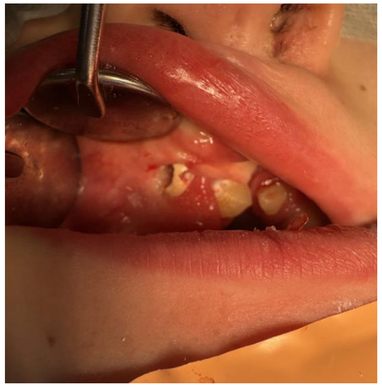
Past Surgical History: 10/2009 Cleft Lip repair, 09/2010 Cleft Palate repair, 02/2016 Dental Restorations and Extractions, 03/2025 Dental Restorations and Extractions under General Anesthesia

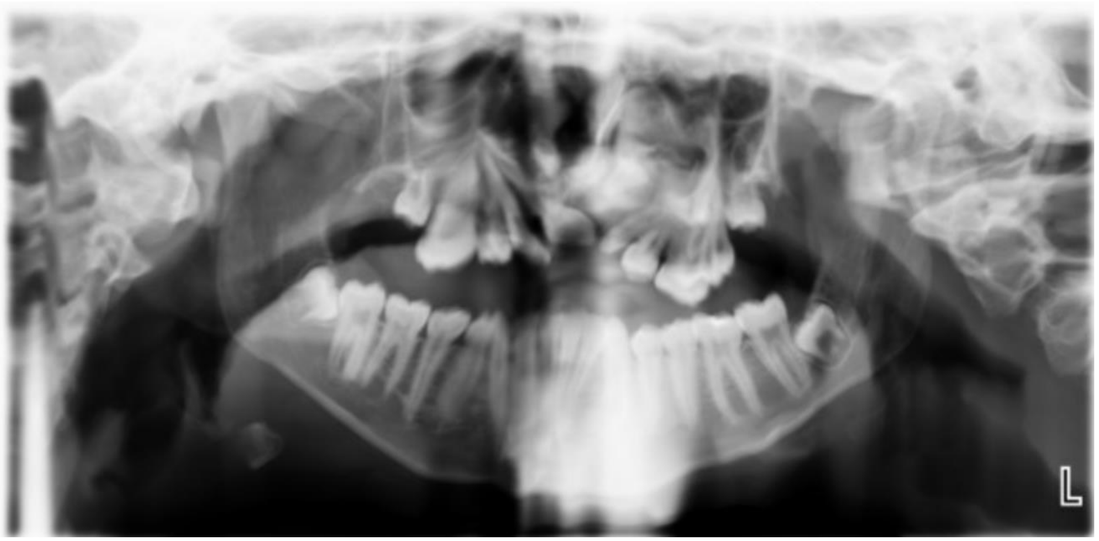
CLINICAL PHOTOS











CLINICAL MANAGEMENT

Other care received in the dental clinic:

- 01/2018, composite restoration on #19-O using a protective stabilizer
- 09/2021, clinical exam revealed large unrestorable caries on tooth #9 and recurrent caries under a failing restoration on tooth #8. Both teeth were extracted.
- 10/2024, a panoramic radiograph showed findings consistent with bilateral cleft palate. The mandibular arch demonstrated age-appropriate growth with developing third molars. Multiple malpositioned were present in the anterior and posterior left maxilla, suggestive of supernumerary or malformed teeth. Severe maxillary crowding
- 03/2025, the patient was treated under general anesthesia. Extraoral exam showed no edema, erythema, or lymphadenopathy. Intraoral findings included a high, narrow palate consistent with cleft repair, poor oral hygiene with generalized moderate plaque, and mild to moderate gingivitis, most pronounced in the maxillary anterior region

A thorough dental cleaning was performed. Eight sealants were placed for prevention, two composite restorations were completed for carious teeth, and a permanent canine toot, #6, located in the cleft area was extracted. Notably, this tooth was only visible upon full retraction of the upper lip.

The patient will continue routine follow-up with the hospital dentistry clinic and the craniofacial anomalies team for ongoing care and management.

REFERNCES

Cleveland Clinic. (2023). Dandy-Walker syndrome. Retrieved April 6, 2025, from https://my.clevelandclinic.org/health/diseases/6002-dandy-walker-syndrome

Dashe, J. S., & Norton, M. E. (2024). Prenatal diagnosis of CNS anomalies other than neural tube defects and ventriculomegaly. UpToDate. Retrieved April 6, 2025, from *Up to Date*

Hart, M. N. , Malamud, N. & Ellis, W. G. (1972). The Dandy-Walker syndrome. Neurology, 22 (8), 771-780.