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Dental considerations and management of a patient with dandy-walker syndrome: A case report

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ABSTRACT

Introduction: Dandy-Walker syndrome is a congenital disorder that mainly affects brain development during pregnancy, particularly the cerebellum, which is the part responsible for movement. An enlargement in the fourth ventricle and cyst formation occurs in this disorder. As cerebrospinal fluid accumulates, high pressure and swelling may occur in the base of the skull causing hydrocephalus. *Objective:* The purpose of this article is to describe the clinical manifestations, dental considerations, oral findings and management strategies in a DWS-affected pediatric patient. *Case description*: 4 years old Saudi male patient came with his father with a complaint of multiple caries. The patient was completely uncooperative and refused to sit on the dental chair for examination. The patient was diagnosed with Dandy-Walker syndrome and presented to the clinic with difficulty in swallowing and chronic carious lesions. The swallowing problem is being directed to the swallowing unit at the speech and language pathology department. Under general anesthesia, the patient will go for full dental rehabilitation. *Conclusion*: Children with Dandy walker syndrome are presented with different clinical presentations and varied intellectual development. The dental management of such children should be costumed for each individual based on their needs. Oral health education for parents and caregivers regarding oral hygiene and dental trauma is of prime importance.

Keywords: Dandy-Walker Syndrome, Hydrocephalus, General Anesthesia, Full Dental Rehabilitation, Pediatrics, Case Report

1. INTRODUCTION

The Dandy-Walker syndrome (DWS) is a congenital malformation of the brain first described by Dandy and Black fan in 1914 (Alexiou et al., 2010). An irregularity in the fluid-filled spaces and cerebellum occurs in this condition (Imataka et al., 2007). It is characterized by an enlargement of the fourth ventricle, a small channel that allows fluid movement in the upper and lower parts of the brain and spinal cord. Agenesis of the cerebellar vermis to a

limited or complete extent, which connects the two cerebellar hemispheres and cyst formation in the posterior cranial fossa (Imataka et al., 2007; Sawaya and Mc Laurin, 1981). In addition, this condition may be complicated by the occurrence of hydrocephalus in almost 80% of the cases (Spennato et al., 2011).

The exact etiology remains unknown. It is believed to be a result of the interaction between environmental and genetic factors. Environmental factors such as prenatal exposure to alcohol, teratogenic agents, certain types of medicines and infections might play a role. Also, maternal diabetes has been reported as a predisposing factor for Dany-Walker malformation (Sawaya and Mc Laurin, 1981). With no clear pattern of inheritance, most of the cases are rare with a limited percentage reported to have a genetic origin (Sawaya and Mc Laurin, 1981). In this malformation, several parts of the cerebellum undergo abnormal development, causing anatomical abnormalities that can be seen with neuroimaging (Ecker et al., 2000). Therefore, the diagnosis of DWS is based on distinctive neuroimaging findings (Ecker et al., 2000) however, a limited number of studies address the DWS dental implications and treatment. In our case report we will describe the dental considerations, clinical manifestations, oral findings and management approaches for a pediatric patient with DWS.

2. CASE PRESENTATION

A 4-year-old Saudi male patient presented with his father to the pediatric dentistry clinic at King Saud Medical City, Riyadh, KSA. He was referred by the pediatric neurology department for dental consultation and management. According to the father and patient's medical record, the child is a known case of Dandy walker syndrome with corpus callosum agenesis, moderate bilateral hydronephrosis, global developmental delay and intellectual disability. The patient is a full-term child with a history of neonatal intensive care unit admission in the first 3 months of his life. He was on a prophylactic antibiotic (Amoxil 50mg once daily) for the first three years of his life for hydronephrosis. His family history was unremarkable for DWS as well as other neurological diseases. The patient was born to non-consanguineous parents with two healthy siblings. He is living with his parents and the mother is the primary caregiver. Also, he is attending special needs school for early intervention and rehabilitation. Dietary history revealed child is on a soft diet only due to difficulty in swallowing and he is following up with the swallowing unit at the speech and language pathology department. No history of night bottle feeding, nor gastrointestinal tube (GT) was found.

Upon clinical examination

In the first dental visit and due to the patient's medical condition, the father mentioned that he had never sought dental treatment for his child before and he was concerned about his decayed front teeth. He was not aware of a history of dental pain due to the child's inability to communicate. On extra-oral examination, the patient presented with short stature, distended fontanels, frontal bossing and the occipital bulge was evident. The orbits were deeply sunken with low-sets quinting eye sand strabismus. The external auricle showed poorly lobulated ears. Ataxic gait as he walks, slurred speech and difficulty in holding things with his hands were observed. These findings with a history of difficulty of swallowing indicate a lack of muscle coordination affecting both gross and fine motor skills. The child was not cooperative to be examined on the dental chair, so an intraoral examination was conducted while the father is holding the child in his lab. The child is in the primary dentition stage, with severe early childhood caries and poor oral hygiene (Figure 1). Soft tissue examination showed all within normal limits with a slightly high palatal vault (Figure 1). His airway assessment was Mallampati classification II. The patient was asymptomatic and there were no clinical signs of dental abscesses or acute infections. According to the American academy of pediatric dentistry caries risk assessment tool, the child is classified as a high caries risk, due to the presence of multiple cavities, visible plaque on teeth and special health care needs. The case was explained to the father and a treatment plan was formulated by starting with the preventive phase of oral hygiene instructions. Based on the child's medical condition and extensive dental treatment needs as well as his inability to cooperate due to his intellectual disability, a decision was made to treat the patient in the operating room setting under general anesthesia. Upon consulting with his medical team, there were no contraindications for general anesthesia. Upon Pre anesthesia assessment by an anesthesiologist and based on the American society of anesthesiologists (ASA) classification, the patient is categorized as ASA II.

Upon radiographic examination

Radiographs were taken while the patient is sedated on the operation table. Right and left Bitewings radiographs showed deep caries of the upper right primary first molar and lower right and left primary first molar. Periapical radiographs revealed three rooted lower primary second molars, a heart-shaped pulp chamber of the lower left primary central incisor and deep caries in upper primary anterior teeth (Figure 2).

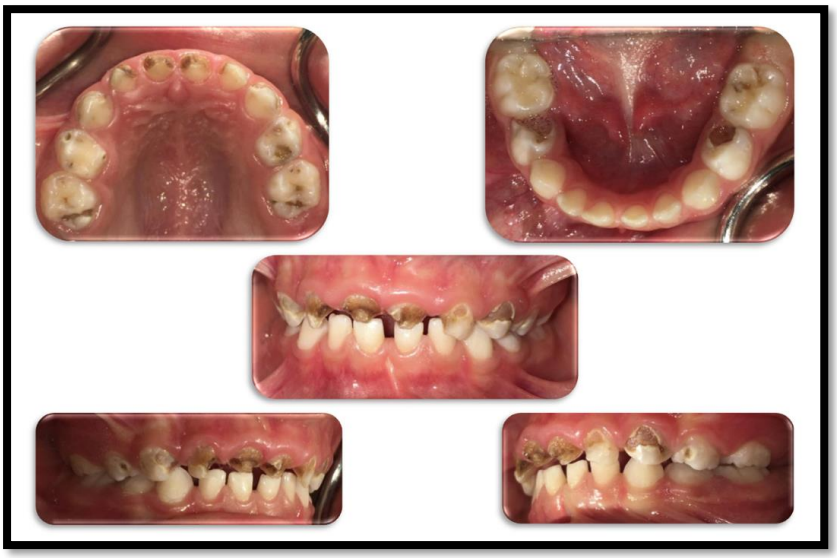

Figure 1 Preoperative intra-oral photographs

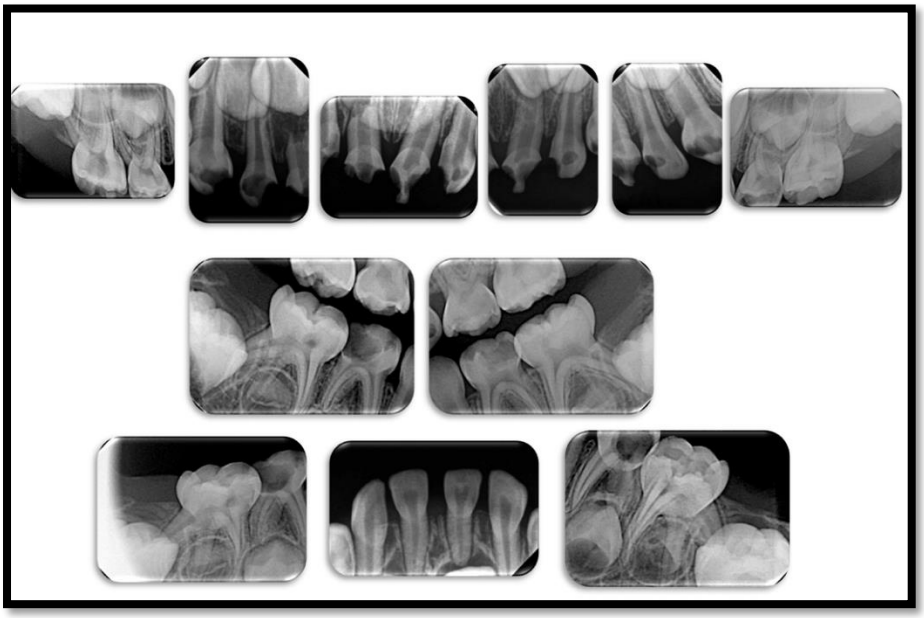

Figure 2 Preoperative intra-oral radiographs

Management

Due to the patient's low cooperation level, an intravenous (IV) line was not inserted; therefore, sevoflurane and oxygen were administered using a facemask. Followed by an IV propofol and endotracheal intubation with the standard laryngoscopic technique was done. To reduce oral edema, dexamethasone was administered and anesthesia was maintained with oxygen-sevoflurane. Vital signs were stable during the operation. With the exception of induction and extubation, the procedure lasted 110 minutes. For the restorative and surgical phase, all upper posterior primary teeth were treated with formocresol pulpotomy and then restored with prefabricated stainless-steel crown SSC (3M ESPE, SSC, Germany). The lower primary second molars were also restored with prefabricated stainless-steel crown SSC (3M ESPE, SSC, Germany). Upper primary canines were restored with light-cured composite resin Prime and adhesive (Adaper Scotchbond™ Multi- purpose, 3M ESPE, St. Paul, MN) was applied and cured according to manufacturer instructions and the cavities were restored with shade A2 of Z100TM composite material (3M ESPE, St. Paul, MN). Whereas upper primary incisors and lower primary first molars were extracted. Afterward, a topical fluoride varnish is applied (Duraphat from Colgate Oral Care, Sydney, Australia) to all teeth. When the operation has been completed, the return to spontaneous breathing was smooth, the endotracheal tube was extubated and administered of 100% oxygen. The dental procedure was successfully conducted and there were no postoperative complications. The patient was discharged from the hospital on the

same day. He was seen in a follow-up visit in the dental clinic two weeks postoperatively, with no post-operative complications reported. For the maintenance phase, the patient was booked after 3 months for a recall visit.

4. DISCUSSION

Dandy-Walker malformation (DWM) is a rare congenital brain malformation that involves the cerebellum. Part of the brain is the cerebellum which regulates movement coordination and because of that, children with DWS have several neurodevelopmental symptoms in addition to, intellectual disability and behavioral disorders (Alexiou et al., 2010; Sawaya and Mc Laurin, 1981; Stambolliu et al., 2017). In early infancy, the symptoms are usually noticeable and observable (Stambolliu et al., 2017). The milder the form of DWS, the fewer neurological symptoms (Sasaki-Adams et al., 2008). Our patient in this case report is considered to have mild DWS since he has no hydrocephalus or seizures. According to studies, craniofacial abnormalities such as cleft lip/palate, high-arched palate and retrognathia are more common among patients with DWS (Imataka et al., 2007; Sawaya and Mc Laurin, 1981). No studies in the literature found a correlation between any dental anomalies and DWS. In our case report, we found that the child has an abnormal pulp chamber shape of the lower left central primary incisor, although no history of dental trauma was stated by the father. Which might be affected by local trauma during intubation in NICU. Also, he has three rooted lower primary second molars which might be a coincidental finding with such syndromes.

Children with DWS have poor fine motor skills which creates a challenge for their oral hygiene habits. Caregivers should keep in mind that those kinds of patients can't brush their own teeth and brush should be done for them. Most DWS children are taking medications for a long period of time which might be a risk factor for developing dental caries. Also, due to their poor muscle coordination, they are borne for more frequent falls and dental trauma. Anticipatory guidance should include such risks. Due to their inability to cooperate, it is difficult to treat them in a regular dental setting. In most cases, general anesthesia is required to treat them. For the previously mentioned reasons, pediatric dentists tend to follow a more aggressive treatment approach utilizing stainless-steel crowns whenever possible.

4. CONCLUSION

Children with Dandy-Walker syndrome show variable clinical presentation and intellectual development. It is critical that the dental management of such children is tailored to the individual's needs. A key part of oral health education for parents and caregivers is teaching them how to care for their children's teeth.

Author Contribution

All authors contributed equally to the manuscript.

Informed consent

Written and oral informed consent was obtained from all individual participants included in the study. Additional informed consent was obtained from all individual participants for whom identifying information is included in this manuscript.

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Conflict of interest

The authors declare that there is no conflict of interests.

Data and materials availability

All data sets collected during this study are available upon reasonable request from the corresponding author.

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