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Dravet Syndrome from Dental View: A Case Report

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ABSTRACT

Dravet Syndrome is a complex neurological disorder characterized by developmental delays and seizures caused by Gingival hyperplasia (gingival enlargement and frequent bleeding) in a paediatric, four-year-old girl for several years. This condition, often exacerbated secondary to medications used to manage DS, contributed to the development of severe dental caries—the girl with severe dental caries and gingival hyperplasia presented to the clinic. A collaborative approach involving pediatric dentistry, neuro-paediatrics, general medicine, and anaesthesia was employed to extract affected primary teeth—this multidisciplinary effort streamlined management and treatment, leading to successful outcomes. Postoperatively, the patient showed satisfactory recovery and underwent regular follow-ups. Regular follow-up appointments conducted every three months demonstrated significant improvement in the girl's oral health and overall well-being, including a positive impact on her emotional, cognitive, and motor functions.

INTRODUCTION

Dravet syndrome (DS) is a rare form of epilepsy; developmental and epileptic encephalopathy typically begins in infancy. (Meskis, 2022) The lifestyle of DS patients is adversely affected, characterized by seizures, developmental delays, behavioural problems, emotional impairment, feeding problems, sleep problems and social isolation. (Stein *et al.*, 2019) Children with DS show slow or no development in cognitive and motor skills. (Satta *et al.*, 2020) Some children and adolescents show externalizing behaviour, aggression, activeness and harmful behaviour. Some adolescents and young adults have been reported to exhibit internalizing behaviours and routinized and compulsive habits. (Inácio, 2023) This syndrome also affects the patient's parents and family, as there is no cure for DS, but treatment and support from family can help manage the symptoms and improve the quality of the patient's life. (Sinoo *et al.*, 2019)

Patients are reported to experience various oral health problems: disruptive and delayed dental development, grinding of teeth, pain and discomfort, usually due to several factors, including motor skills and coordination, epileptic seizures, oral motor skills and as a secondary to medications used to treat Dravet Syndrome. These complications can lead to nutrition deficiency, gait abnormalities, and muscle functions such as difficulty chewing and speaking; thus, overall health is affected. Therefore, such a patient presents a multifaceted threat to dental care: uncontrolled seizures, heightened pain sensitivity and potential behavioural challenges to smooth execution of dental diagnostics and treatment.

This case report highlights a dental view of pediatric patients with DS. It includes dental diagnosis and treatment, and it later impacts the quality of life of a patient.

LITERATURE REVIEW

The uncommon and severe form of epilepsy known as Dravet Syndrome poses considerable complications for dental therapy because of its correlation with cognitive deficits, behavioral abnormalities, and drug-related adverse effects (Sinoo *et al.*, 2019). While the neurological aspects of DS have been extensively studied, its dental implications remain relatively understudied in the literature. Limited research suggests that individuals with DS may present unique dental challenges and oral health considerations. For instance, prolonged or frequent seizures, common in DS, may increase the risk of dental trauma, including tooth fractures or avulsion (Slayton *et al.*, 2020). Currently, little research is available on the oral effects of Dravet syndrome, emphasizing the value of case studies in clarifying clinical presentations and treatment plans. Prior research has indicated that people with epilepsy are more likely to experience oral health problems, such as dental caries, periodontal disease, and malocclusions (Bakaev, 2023).

Additionally, gastrointestinal and eating difficulties have been reported to be prevalent in Dravet Syndrome Patients, which poses a burden on caregivers and food management. Thus emphasizing the importance of nutritional challenges caused by eating habits (Minderhoud *et al.*, 2023). Oral care regimens are further complicated by the development of gingival hyperplasia and xerostomia, which have been linked to the use of antiepileptic medications like phenytoin and valproic acid. Moreover, Comprehending the distinct obstacles presented by Dravet Syndrome from a dental standpoint is crucial to guaranteeing all-encompassing and customized treatment strategies. Further research in this area is needed to elucidate the specific oral health needs of individuals with DS and to develop tailored dental management

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protocols to improve their oral health outcomes. As the dental profession continues to recognize the importance of interdisciplinary collaboration in managing complex medical conditions, including epilepsy, efforts to integrate dental care into the comprehensive management of DS patients are essential to optimize their overall health and quality of life.

METHODS

Patient's Information

A Darvet syndromic four-year-old girl appeared at a dentist clinic on January 10, 2021. She had a major complaint of severe dental caries on her primary teeth, causing pain and intra and extra-oral swelling. Previously, the patient was reported to have gingival enlargement for a few years, causing malocclusion, difficulty in chewing and frequent gingival bleeding. Despite these problems, the patient always had regular dental checkups and cleanings.

Physical Examination

The patient's face was asymmetrical and seemed swollen from the region of the lower jawline, chin and cheek.

Clinical and Radiographic Examination

Extra orally, both the right and left sides of the submandibular region were swollen and tender. Intra-orally, several dental concerns were noticed, including Generalized severe dental caries, erythematous (inflammation), bleeding gums, and no signs of periodontal disease. Radiographs of the patient's teeth showed caries on primary teeth with radiolucent lesions (abnormal tissue, i.e. enlargement of gums) on the furcation area (roots of the multi-rooted tooth branch out). No bone loss or periapical pathology was found.

RESULTS

Figures 1, 2 and 3 are evidence of pre-operative X-rays scanned in 2021.

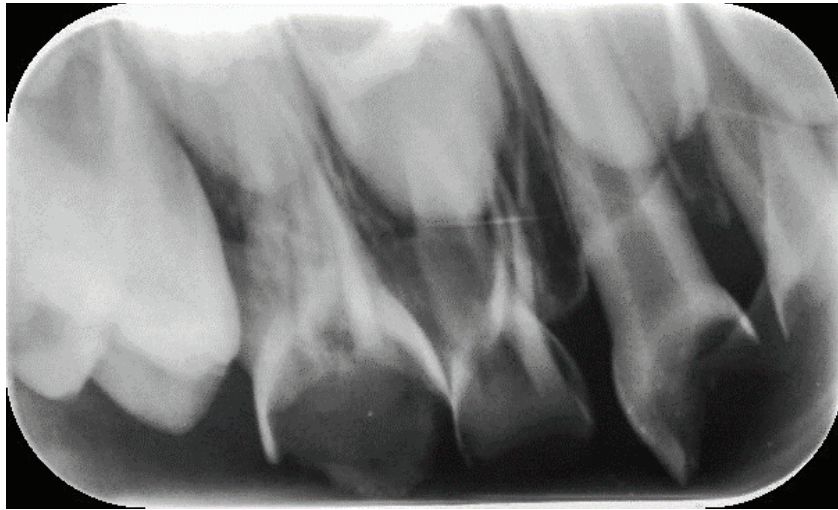


Figure 1: Pre-operative x-ray (2021)

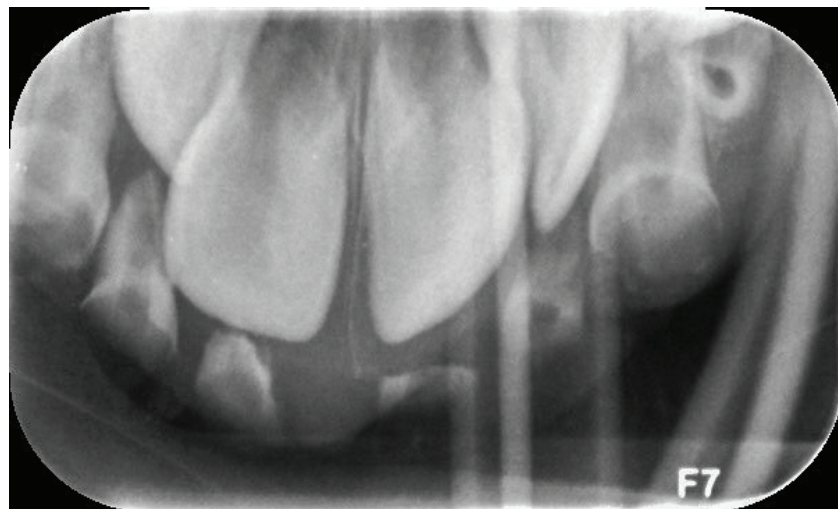


Figure 2: Pre-operate x-ray (2021)

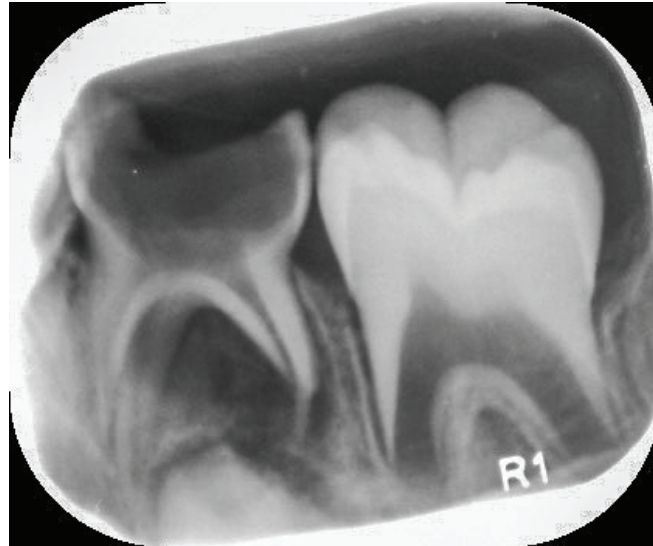


Figure 3: Pre-operative x-ray (2021)

Diagnosis

Gingival hyperplasia and dental caries occurred as a potential complication due to Dravet Syndrome’s medication side effects.

Treatment Plan and Method

Due to complexities, the patient was referred to a collaborative approach of a pediatric dentist, neuro-pediatric, general and anaesthesia teams. They assisted the pediatric dentist in providing a dental treatment, which included pulp therapy, composite restoration, crowning for primary and permanent teeth, and extraction of defected primary teeth.

While ensuring painless treatment to extract all the affected primary teeth and managing uncontrollable seizures and behaviour, the pediatric patient booked an appointment to undergo general anaesthesia on January 27, 2021, for the procedure. After getting clearance from the collaborative team, the patient was admitted to the hospital a day before the operation. During treatment, the patient was placed in a supine position, nasal intubation was performed, all primary teeth were extracted, and bleeding was controlled. The child was sent to the ward, where she was kept under observation. Once stable, she was discharged by a general paediatrician the other day after the operation.

Follow-up and Outcome

A postoperative follow-up visit was scheduled after two weeks, and a satisfied patient’s health status was observed. Further, the child was kept under observation every three months until 2024.

First Follow-up Visit

The child had her lower molars erupted, and some caries were noticed, which were restored at the clinic.

Second Follow-up Visit

The child’s permanent teeth erupted earlier than usual.

Third Follow-up Visit

The girl had a smile on her face. She was seen recognizing the clinic and her dentist. Moreover, the patient’s mother reported that the child’s motor action was improved as she asked neuro-pediatric to discontinue medication for Dravet syndrome.



Figure 4: Follow-up evidence (2024)



Figure 5: Follow-up evidence (2024)



Figure 6: Follow-up evidence (2024)

Figures 4, 5, and 6 show sound tooth structure; there are no caries, and the restored primary tooth remained satisfactory.

DISCUSSION

Dravet Syndrome is a rare type of epilepsy that occurs due to SCN1A gene mutation, and key characteristics are seizure onset in infancy, developmental delay, and behavioural problems (Clayton *et al.*, 2022). It affects various regions of the brain, specifically the hippocampus (involved in epilepsy), prefrontal cortex and hippocampal dentate gyrus (regions associated with emotions and memory). Thus, the interplay of cognition, emotional impairments and behavioural patterns contributes to comorbidities as a salient feature of this syndrome (Makiello *et al.*, 2023). Common examples include muscle impairment, sleep problems, and gait impairment; among these, feeding difficulties and weight loss are frequently reported (Meskis, 2022). Despite the prevalence of feeding difficulties, the underlying factors are still in pipe of exploration. On the other hand, dental problems are becoming popular and common among DS individuals primarily due to frequent seizures, which, if uncontrollable, can damage teeth and gums (Madaan & Sahu, 2022).

Individuals with Dravet Syndrome may experience various oral effects, gingival hyperplasia and submandibular swelling should be regarded as secondary concerns, given the significant implications for the submandibular region in saliva production, sensory and motor function, and lymphatic drainage (Zapoozhets *et al.*, 2019). These conditions can influence oral issues such as tooth decay, dry mouth, difficulty swallowing, and speech problems. Moreover, orofacial dyspraxia can be challenging for DS individuals to maintain a simple oral hygiene regime, such as brushing or flossing teeth (Minderhoud *et al.*, 2023).

It is highly recommended to have early interventions, regular dental checkups and cleanings to prevent serious dental complications and facilitate individuals to have a better life (Inácio, 2023). Furthermore, individuals should know the signs and symptoms that cause dental complications.

In addition to the oral manifestations previously discussed, there are further considerations regarding the dental health of individuals with Dravet syndrome.

The process of teeth eruption itself may present unique challenges, chewing, bite and speech problems and gingival hyperplasia frequently complicating this normal developmental stage. These factors underscore the importance of tailored dental care strategies for individuals with Dravet syndrome to mitigate associated risks and promote optimal oral health outcomes.

CONCLUSION

In conclusion, this case report highlights the challenges and successful diagnosis and treatment of a pediatric patient suffering from severe dental caries on primary teeth along with intra and extra-oral swelling, causing overall pain and discomfort with Dravet Syndrome. Implementing a collaborative approach to parents' continuous support, integrated with the thorough dental history of a patient, led to favourable outcomes. It proved to be an emerging dental treatment of a DS patient, yet the patient's regular checkups and timely actions were instrumental. The permanent teeth erupted earlier than usual time. The girl's smile and recognition of her dentist and clinic indicated permanent teeth erupted than earlier, improved emotional health and cognition. Parents also reported improved performance and motor skills of the child at school. Overall, the case highlights the importance of dental care in overcoming dental challenges in Dravet Syndrome patients.

Future Recommendation

Further research can explore different dental dimensions of Dravet Syndrome pediatric patients; a database can be collected to develop awareness for common signs and symptoms, tackle other interconnected factors which arise due to dental problems, assess oral health complications, and develop targeted preventive measures that can come into existence.

This case report aims to enhance understanding and treatment of Dravet Syndrome concerning dental health, contributing to medical knowledge and emphasizing the significance of tailored dental approaches for optimal patient outcomes.

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