Dental Treatment Following Trauma in Patient with Dravet Syndrome: A Case Study

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INTRODUCTION

Dravet Syndrome (DS) is a rare form of epilepsy that begins in infancy or early childhood and can include a spectrum of symptoms ranging from mild to severe. These symptoms include clonic or tonic-clonic protracted seizures, developmental disabilities, motor skill difficulties, hyperactivity, communication difficulties, and behavioral abnormalities. Other symptoms can occur, including sleep disorders, infection susceptibility, affected growth, and forward-leaning, stiff walking. Most cases of DS are caused by a mutation in the SCN1A gene found on chromosome 2 (2q24.1). Children with DS can have delayed eruption of teeth and a small number can experience dental developmental disruptions, which has resulted in teeth of smaller size or different shape. Medications used in the treatment of DS can lead to dry mouth, heartburn, vomiting, and increased bleeding tendency. As with other seizure disorders, patients are at an increased risk of dental injury and any necessary treatment for these injuries is affected by the patient's ability to tolerate it. Often times, sedation or general anesthesia must be considered.

CASE REPORT

A 14-year-old female presented to the dental clinic at Rainbow Babies and Children's Hospital on May 21, 2024. The patient has a past medical history of DS, carnitine deficiency due to valproate use, hypotonia, cognitive deficit and developmental delay (patient has cognitive capacity similar to 3-year-old, per parents). Medications: clobazam, clonazepam, cyproheptadine, diazepam, epidiolex, levocarnitine, melatonin, ondansetron, stiripentol, and valproate. The patient had experienced a seizure on May 12, during which she fell and struck her front teeth on the pavement. Patient was taken to another hospital system's emergency department and was treated for other concerns and was told to call our office for dental follow-up. Clinical and radiographic exam was completed, with the following findings: well-healing soft tissue around 8/9 in the frenum area with a bit of tissue right in between 8 and 9 still healing. No mobility associated with 8 or 9. Tooth 8 and 9 appear slightly out of position compared to the rest of arch. No trauma to remaining dentition. Maxillary occlusal radiograph taken and all radiographic findings within normal limits. Differential diagnosis: #8: lateral luxation, #9: lateral luxation. Tooth 8 and 9 were noted to have abnormally widened canals and closed apices; patient showed slight delay in exfoliation of primary teeth. No other caries noted. Plan was made to monitor the teeth 8 and 9 for signs and symptoms of infection. Patient returned on May 28, 2024—parents stated that although patient had not communicated any pain, patient had been touching around the area of 8/9 and around her nose. Exam and radiographs revealed possible radiolucency on #8. Given the signs and symptoms of pain/discomfort and inability to gain ample information from patient regarding complete diagnosis, it was recommended to see patient under general anesthesia for further evaluation of #8 and 9, followed by treatment, as needed.

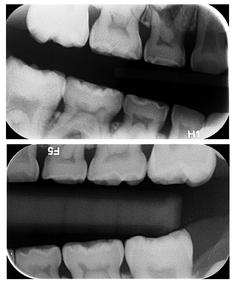
CLINICAL AND RADIOGRAPHIC PRESENTATION

Clinical photo (5/12/24)

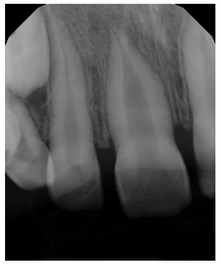
Initial (5/21/24)

Follow up (5/28/24)

Bitewings (5/28/24)



Initial on DOS (9/19/24)



Completed

SUMMARY OF TREATMENT RENDERED

Pulpal testing on day of surgery (during pre-operatory):
Tooth #8:

- Cold testing: Negative, no response or movement from patient
- Palpation: within normal limits, patient showed little reaction
- Percussion: patient moved head back quickly after percussion testing

Tooth #9:

- Cold testing: Positive, patient moved away slightly but quickly recovered
- Palpation: within normal limits
- Percussion: within normal limits

Treatment rendered under general anesthesia:

- Endodontic therapy #8:
    - Canal was accessed using high speed bur, coronal area was cleaned and shaped using Endo Z and Gates-Glidden burs – **pulpal tissue was deemed to be necrotic.**
    - Canal was cleaned and shaped with rotary files and irrigated thoroughly
    - Obturation was achieved by placing **60/.04** gutta percha file to working length of 22.5 mm. Accessory cones placed: **80/.04, 40/.04, 40/.04, 40/.04, 40/.04** – cold lateral condensation performed between each cone.

DISCUSSION AND CONCLUSION

Dravet syndrome (DS) presents many challenges to the patients who struggle with it and their families. Dental care for these patients is one of these challenges. This patient presented with a fairly common injury for children generally, however, her care was complicated by several factors related to her condition. The injury was sustained during a seizure episode caused by DS and the treatment needed required general anesthesia due to her developmental delay. Although no definitive link has been demonstrated between patients with DS and abnormally shaped teeth, there have been reports of "small" or "different" shaped teeth, thought to be related to developmental disruptions for these patients. For this patient, it was certainly the case that her central incisors had developed abnormally, which necessitated a unique obturation.

During post-operative discussions with father, the recommended follow-up schedule was reviewed to monitor the completed root canal on tooth #8. It was also noted that patient has delayed eruption of primary teeth which could also be related to a developmental delay. Father was informed that patient shows evidence of bruxism, which he confirmed and asked if any treatment would be possible. Due to the patient's questionable ability to comply with an appliance and since the patient is in a time period of facial growth and tooth exfoliation, it was not recommended that we fabricate an occlusal guard at this time. Instead, it was suggested that the father could buy a traditional "boil-and-bite" mouth guard to test patient compliance and provide a more economical option.

This patient's case provides a prime example of treating dental trauma and its sequelae for patients with special health care needs, especially those who are prone to trauma due to their condition and/or have difficulty expressing pain, discomfort, or describing symptoms. Providing the correct diagnosis and treatment amid these other concerns is a vital role for the pediatric dentist.

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