



ABSTRACT/INTRODUCTION

Tooth development anomalies can happen at many stages of cell development and are relatively common. These anomalies can be isolated findings however, they may also indicate minor or major associated syndromes and identification of these findings are essential to providing comprehensive care for patients. Dentin Dysplasia Type II is a rare autosomal dominant disease resulting from a disruption in the apposition stage of cell development. This disease affects the primary teeth and clinical manifestations can include bulbous crowns, cervical constriction, mild discoloration (amber color) and pulp obliteration. Permanent teeth tend to look typical but can demonstrate thistle-tube shaped pulps and/or multiple pulp stones. This case presentation discusses an 8-year-old male patient who presented with chronic decay and enlarged pulps noted at first clinical exam and radiographic interpretation. Following restorative treatment, the patient experienced loss of Stainless-Steel Crown (SSC) with remaining coronal tooth structure due to pulpal obliteration. A differential diagnosis of Dentin Dysplasia Type II was determined with the aid of a local oral pathologist. Patient was advised to seek genetic testing to improve treatment planning capabilities and further recommendations.

CASE REPORT



[Figure 1](#)

An 8 year- 6 month old male patient presented to the Children's Mercy Hospital (CMH) dental clinic for a new patient exam 4/12/21. Medical and dental history was reviewed with child's mother and no significant medical history was determined. Clinical and radiographic examination revealed mixed dentition, hypoplastic 1st permanent molars and incisors, generalized moderate caries, idiopathic internal resorption on #B with no symptoms, and moderate plaque buildup (Figure 2). Restorative care was completed first in the lower right quadrant and included Stainless Steel Crowns (SSC) #S/#T and an occlusal buccal composite #30.

Patient then presented for a limited oral exam in November of the same year due to loss of crown #S (Figure 1). Radiographs revealed pulpal obliteration of tooth structure #S with minimal mesial root remnants (Figure 3). Further evaluation utilizing CBCT and panoramic radiographic imaging revealed teeth #A, B, C, H, I, L and R to have enlarged pulpal chambers (Figure 3 & 4). An oral pathologist was consulted regarding idiopathic internal root resorption and a preliminary diagnosis of Dentin Dysplasia Type II was suggested. The family was encouraged to seek genetic counseling for definitive diagnosis to aid in further treatment planning. Completion of dental caries control was encouraged.

[Figure 2](#) BW 4/12/21



[Figure 3](#) BW 11/12/21



[Figure 3](#)



[Figure 4](#)

DISCUSSION/CONCLUSION

Dentin Dysplasia type II is a rare autosomal dominant disorder that is caused by a disruption in the sialoprotein that is required for normal dentin development. This disease can affect the primary and permanent teeth so early diagnosis of this developmental anomaly is essential for appropriate comprehensive treatment planning. Genetic testing can reveal disruption of the sialophosphoprotein gene which is located on chromosome 4 (4q21.3). This protein is important for coding the two collagenous proteins of the dentin matrix and is required for normal dentin formation.¹ Early diagnosis may allow for orthograde endodontics to be considered as a treatment option for permanent teeth to prevent full pulpal obliteration. Routine dental care and adequate oral hygiene are essential keys to prevent complicated or invasive procedures. In conclusion, patients with Dentin Dysplasia type II or other tooth anomalies should receive routine dental care to monitor progression through primary and permanent dentition and genetic testing can provide definitive diagnosis.³

REFERENCES

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