Dental Eruption Pattern in Patients with Congenital Adrenal Hyperplasia

Northwell[™] Staten Island University Hospital

Firoozeh Shekari DDS Staten Island University Hospital

INTRODUCTION

Congenital Adrenal hyperplasia (CAH) is an inherited genetic disorder that results in reduced production of cortisol. This occurs due to the diminished activity of 21-hydroxylase enzyme, which plays a crucial role in cortisol synthesis. Consequently, the adrenal cortex experiences chronic overstimulation. Accelerated tooth eruption has been reported in patients with CAH due to elevated hormonal secretion and prolonged use of corticosteroid therapy.

CASE DESCRIPTION

A10-year-old Hispanic female, who has been a patient of Staten Island University Hospital dental clinic presented for a routine visit. Clinical and radiographic exam revealed early exfoliation of primary teeth and accelerated eruption of permanent teeth. Patient presented with primary molars and canines during a hygiene visit in 2021. However, during a recall visit in 2023 all primary teeth, except maxillary canines, were found to have been exfoliated in less than 2 years. This is considered early exfoliation of primary teeth that is not expected to be seen in a 10-year-old patient. Incomplete root formation was also noted on posterior teeth potentially due to premature eruption of teeth into the oral cavity. Patient was referred for an endocrinology consultation to assess hormone levels, specifically cortisol production. Insufficient activity of 21-hydroxylase enzyme was found leading to diagnosis of congenital adrenal hyperplasia.



Figure 1: Panoramic radiograph in 2021 showing presence of primary molars and canines



Figure 2: Panoramic radiograph in 2023 showing exfoliated maxillary and mandibular molars as well as mandibular canines. Premature eruption of some teeth noted

DISCUSSION AND CONCLUSION

Observations such as premature exfoliation of primary teeth and accelerated eruption of permanent teeth, along with other oral manifestations of CAH can assist the dental provider to make appropriate referrals to medical specialists with the aim to improve patient's overall health. Premature loss of primary teeth associated with CAH has been reported in another case report in the past. Bone destruction has been found to have some correlation with adrenal gland hormone changes, that can explain early bone destruction and tooth exfoliation in these patients. Early exfoliation of the primary dentition is not a common finding amongst children but when it occurs, a systemic disease may be the culprit, for which the early diagnosis can be beneficial.

References

- Di Cosola M, Spirito F, Zhurakivska K, Nocini R, Lovero R, Sembronio S, Santacroce L, Brauner E, Storto G, Lo Muzio L, Cazzolla AP. Congenital adrenal hyperplasia. Role of dentist in early diagnosis. Open Med (Wars). 2022 Oct 27;17(1):1699-1704. doi: 10.1515/med-2022-0524. PMID: 36382053; PMCID: PMC9616050.
- Claahsen-van der Grinten HL, Speiser PW, Ahmed SF, Arlt W, Auchus RJ, Falhammar H, Flück CE, Guasti L, Huebner A, Kortmann BBM, Krone N, Merke DP, Miller WL, Nordenström A, Reisch N, Sandberg DE, Stikkelbroeck NMML, Touraine P, Utari A, Wudy SA, White PC. Congenital Adrenal Hyperplasia-Current Insights in Pathophysiology, Diagnostics, and Management. Endocr Rev. 2022 Jan 12;43(1):91-159. doi: 10.1210/endrev/bnab016. PMID: 33961029; PMCID: PMC8755999.
- Angelopoulou MV, Kontogiorgos E, Emmanouil D. Congenital adrenal hyperplasia: a case report with premature teeth exfoliation and bone resorption. Pediatrics. 2015 Jun;135(6):e1524-9. doi: 10.1542/peds.2014-3577. PMID: 26009625.