

ACCELERATED DENTAL DEVELOPMENT AS A PRESENTING SYMPTOM OF 21-HYDROXYLASE DEFICIENT NON-CLASSIC CONGENITAL ADRENAL HYPERPLASIA

S. Singer¹, O. Pinhas-Hamiel¹, E. Botzer²

¹*Maccabi Health Care Services, Ramat-Hasharon*, ²*Pediatric Dentistry,*

Tel Aviv Sourasky Medical Center, Tel Aviv, Israel

singeros@nonstop.net.il

Introduction: The role of dental age as an indicator of physical development remains controversial. Several investigators found no significant relationship between dental development and other maturity indicators, while others, claim that canine calcification could serve as a useful tool for evaluating skeletal maturation and somatic maturity. Case presentation: A healthy 4.5-year-old girl had dental examination because of early shedding of her teeth. Dental examination revealed an erupting permanent mandibular central incisor. Intra-oral radiographs demonstrated dental age of 6 years. The child was from Ashkenazi origin. Physical examination was normal, height 109 cm (50th %ile); weight 18 kg (35th %ile); Tanner stage I breasts, a normal clitoris with a few coarse dark pubic hairs. Laboratory tests; testosterone 1.7 nmol/l (normal < 0.14); 17-OH-progesterone 21.9 nmol/l (normal < 2.7); androstenedione 3.85 ng/ml (normal < 0.52); DHEA-S 1.1 mmol/l (normal <1.7); undetectable compound S and 17-OH pregnenolone levels. Bone age was equivalent to 7 years. ACTH test showed 17-OH progesterone level > 75 nmol/l at 60' consistent with the diagnosis of 21-hydroxylase deficient non-classic adrenal hyperplasia. Replacement therapy with hydrocortisone 5 mg daily was instituted. Discussion: Mean age of eruption of first permanent tooth in girls is 5.7±0.52 years. Therefore eruption at age of 4.5 years with advanced dental age suggested a systemic condition. While advanced bone age and precocious puberty are well-known manifestations of non-classic congenital adrenal hyperplasia, this is the first report of a diagnosis based on accelerated dental age.

