
Microdontia, hypodontia, short bulbous roots and root canals with strabismus, short stature, and borderline mentality

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A case of unusual dental findings associated with mild growth and mental retardation is presented. The patient, a 15-year-old girl, manifests small teeth with peculiar short bulbous roots and roundly widened root canals, congenital absence of permanent teeth, strabismus, short stature, and borderline mentality.

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Short tooth roots in association with short stature has been described in five members of a large family,¹ in two similar sporadic cases with microcephaly,^{2,3} and in patients with Rothmund-Thomson syndrome⁴ and bird-headed dwarfism of Seckel.⁵ However, in none of these cases—the reported ones with short roots without concomitant short stature⁶⁻⁸ or any of those with known defects of tooth size and shape⁹⁻¹¹—have the patients had small permanent teeth with bulbous roots and widened root canals that exhibited ovoid dilatations. A sporadic case with this tooth morphology in association with hypodontia, strabismus, short stature, and borderline mentality is presented.

CASE REPORT

A 15-year-old girl was referred to the School of Dentistry because of unusual tooth morphology, which was accidentally revealed during an orthodontic screening program of the Department of Health and Social Security. According

to her father, the girl was born after an uneventful, full-term pregnancy with no exposure to radiation or any unusual agent. She was of normal length and weighed 3.600 gm at birth. Her psychomotor milestones were delayed. She was hospitalized at the ages of 3 and 11 because of failure to thrive, at the age of 5 for surgical correction of strabismus, and at the age of 14 because of bronchial asthma. Menstruation started at 14 years. Eruption of the primary dentition started at the age of 9 months and the permanent dentition at 7 years. No previous radiographs of the teeth were available.

Her parents are nonconsanguineous. The 52-year-old father was examined and appeared normal, although his height, 163 cm, is in the third percentile of height for the country's population. The 43-year-old mother, the 12-year-old brother, and the 7-year-old sister were reported to be normally developed and appeared so in their photographs. Panoramic radiographs of the family members did not exhibit any similarity to the patient's dentition.

On physical examination, the girl appeared short and thin, with a well-proportioned body. Height (149.5 cm), weight (37 kg), and head circumference (51.5 cm) were all below the third percentile. In addition, she had slight strabismus, nevi on the face, and normal pubertal development. Oral examination found 24 small, caries-free tooth crowns, discoloration and nonvitality (from a trauma 3 years earlier) of the maxillary right central incisor and extreme mobility of the ipsilateral lateral incisor, occlusal attrition of all the four central incisors, slight mobility of all teeth, slight generalized marginal gingival hyperplasia, and otherwise normal mucosa.

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Fig. 1. Panoramic radiograph of patient at 15 years of age. Note small permanent teeth with short bulbous roots and roundly widened root canals; absence of permanent maxillary lateral incisors, mandibular second premolars and second molars, and four third molars; retention of primary maxillary lateral incisors and right second molar, and both mandibular second molars. Primary teeth do not manifest bulbous roots.

Measurements of clinical tooth crowns were taken on diagnostic casts to an accuracy of 0.1 mm with sliding calipers. The largest mesiodistal crown diameters (LMCDs) of most permanent teeth ranged from -1 SD to -2 SD for normal permanent female teeth.¹² The LMCD of teeth nos. 11, 21, 31, 32, 44 deviated slightly from normal (-0.2 SD to -0.7 SD), whereas those of the four permanent canines and the maxillary right second premolar were relatively small (values ranged from -2.5 SD to -4 SD). The LMCDs of primary mandibular second molars were also smaller (-2 SD and -3 SD), but the LMCDs of the primary lateral incisors were larger than normal¹² (3 SD and 3.5 SD).

Endocrinologic evaluation showed a peak growth hormone value post L-dopa > 20 ng/ml, and normal thyroxin, cortisol, luteinizing hormone, and follicle-stimulating hormone values. Electrocardiogram, small intestinal biopsy, and sweat test were normal. G-banded chromosome analysis revealed a normal female karyotype (46,XX). Psychometrics demonstrated a borderline mentality (IQ 85). Radiographic examination showed anteroposterior diameter of the cranium of 17.5 cm (SD 16.70 to 22.00 cm) and a bone age of 13.3.

A panoramic radiograph (Fig. 1) revealed a distinctive appearance of the teeth. The girl had small permanent teeth with peculiar short bulbous roots and widened root canals exhibiting ovoid dilatations. Ten permanent teeth were congenitally missing—maxillary lateral incisors, mandibular second premolars and second molars, and four third molars. There was retention of five primary teeth without bulbous roots—the maxillary lateral incisors and right second molar and the mandibular second molars. The permanent maxillary right second premolar was impacted (it erupted after 18 months). The permanent maxillary second molars lacked any root formation and had not emerged.

Panoramic radiographs of the parents and two siblings

did not show any similarity to the dentition of the patient. Her father appears to have occlusal attrition of the anterior teeth and slightly short roots of maxillary teeth and mandibular incisors, and her sister has a mandibular incisor missing.

DISCUSSION

Short tooth roots associated with short stature, inherited as a dominant trait, has been described in five members of a three-generation kindred by Witkop and Jaspers.¹ As in our case, none of the affected members had any endocrinologic problem connected with their short stature, and one of them did not have unusual primary molars. In contrast to our case, they had normal skeletal development and intelligence; no teeth were congenitally missing, and the morphology of their short, thin, dilacerated tooth roots differed entirely from our case.

Short roots of the anterior teeth with taurodontism of the posterior permanent teeth associated with short stature and small head have been described in two similar sporadic cases.^{2,3} Short conical roots of permanent teeth have been reported in a patient of short stature with Rothmund-Thomson syndrome,⁴ and in two Japanese brothers with Seckel microcephalic dwarfism.⁵ Diminished root formation, generalized^{6,7} or restricted to the anterior teeth and accompanied by taurodontia and multiple dens invaginatus,⁸ as well as a prominent feature of dentin dysplasia type I,¹³ has also been described.

None of the published cases manifests the peculiar morphology of the roots and root canals and the microdontia and hypodontia found in our case. The association of these characteristic dental features with mild growth and mental retardation and strabismus in our case might well be coincidental or, as we believe, it could represent a syndrome. To our knowledge the association of these specific traits has not been reported previously.

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