

“Otodontal” dysplasia

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The case of a 3½-year-old Chinese boy with the dental abnormalities of “otodontal” dysplasia is reported. Hearing was normal. Dental anomalies consisted of delayed eruption of globe-shaped molars, bulbous deciduous canines, and double pulp chambers in the molars. Radiographs taken 4 years later showed taurodontic molars, supernumerary microdontic teeth, retarded formation of premolars, and probable aplasia of the mandibular second premolars.

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In 1972 Levin and Jorgenson¹ reported a kindred of Italian ancestry with multiple dental anomalies and associated sensorineural hearing loss. The disorder segregated as an autosomal dominant. The authors first coined the term *familial otodontodysplasia*,¹ then *otodontal dysplasia*^{2,3} for this condition. The dental manifestations of the syndrome consisted of large, globe-shaped molars, absent or small premolars, and delayed eruption of affected teeth. However, deciduous and permanent incisors were normal in shape and size. High-frequency hearing loss was detected by pure-tone air-conduction audiometry in individuals with the dental abnormalities. The age at onset of hearing loss varied from early childhood to middle age.

Subsequently Gundlach and coauthors⁴⁻⁶ reported another affected family. They proposed the terms *globodontia* for the globe-shaped dental deformity and *otodontal syndrome* for the concurrence of the dental and hearing anomalies.

Review of the literature revealed reports of otodontal dysplasia in five kindreds, each with several affected individuals, and in two sporadic cases¹⁻¹³; all were in Westerners. We report the case of a Chinese boy with otodontal dysplasia who had normal hearing. It is the first reported case of this syndrome in an Oriental.

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Fig. 1. Dental cast of mandibular dentition of patient at age 3½ years. Deciduous canine was bulbous, with groove on lingual surface. Deciduous first molar was globe shaped, with a pit in central region of occlusal surface; many developmental grooves radiated from this pit and divided crown into lobules of different sizes. Erupting deciduous second molar had just perforated the overlying gingiva (arrows).

CASE REPORT

This 3½-year-old Chinese boy was first seen at the Dental Clinic of Kaohsiung Medical College because of delayed eruption of his posterior teeth. The boy was the product of an uncomplicated, full-term pregnancy for a 30-year-old couple. Birth weight was 3.1 kg. During pregnancy his mother had no illnesses and took no medications. Except for the dental problem, the boy's development was normal.

His first tooth, a maxillary central incisor, erupted at age 4 months. At the time of oral examination the



Fig. 2. Deciduous incisors are normal in shape. Each deciduous canine shows a spotlike defect on labial surface (*arrows*).

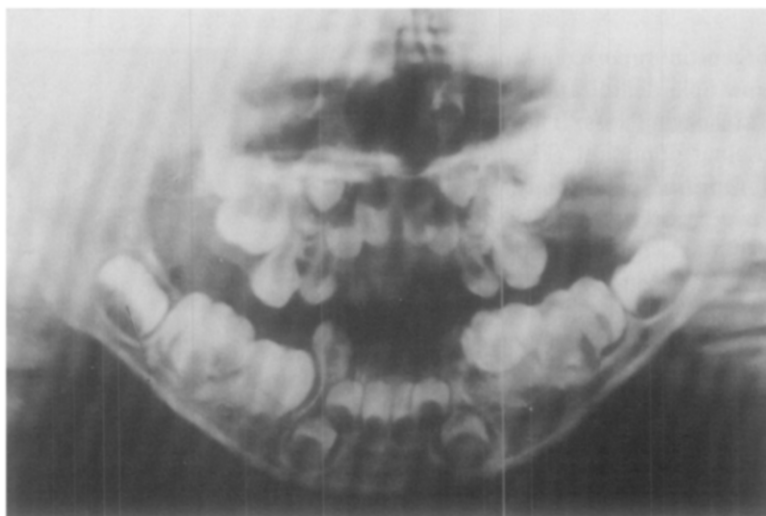


Fig. 3. Panoramic radiograph of jaws at age 3^{11/12} years shows enormous crowns and apparent duplicated pulp chambers of deciduous molars.

deciduous mandibular right first molars were not yet erupted. The deciduous mandibular second molars were just erupting and penetrating the overlying gingiva (Fig. 1). Alignment of erupted teeth was normal. Structure of the incisors was normal (Fig. 2). However, the canines and molars were markedly deformed and large (Fig. 1). Canines were bulbous, and each had a vertical groove extending from the cusp tip onto the lingual surface. Each canine had an enamel defect, 1.5 × 1.5 mm in size, on the cervical third of the labial surface (Fig. 2).

Erupted molars were bulbous. A pit was located in the center of the occlusal surface of each molar; many developmental grooves radiated from these pits onto the buccal, lingual, and proximal surfaces toward the cervical area, and divided each crown into lobules of different sizes (Fig.

1). Radiographs of the patient's jaws showed that all developing deciduous molars had enormous crowns (Fig 3). Each of the crowns of the deciduous mandibular second molars appeared to be either duplicated or fused with a supernumerary component into a twin crown. The primordia of the mandibular premolars, the maxillary second premolars, and the permanent second molars were undiscernible.

The boy was reexamined at age 7^{10/12} years. His deciduous molars were reported to have fully erupted by age 6 years. His face was normal and symmetric (Fig. 4). His hair was straight and clinically appeared of normal density. The permanent maxillary central incisors and the permanent mandibular incisors had replaced their deciduous predecessors. Nevertheless, none of the permanent first



Fig. 4. Patient at age 7½ years. His face was normal and symmetric.

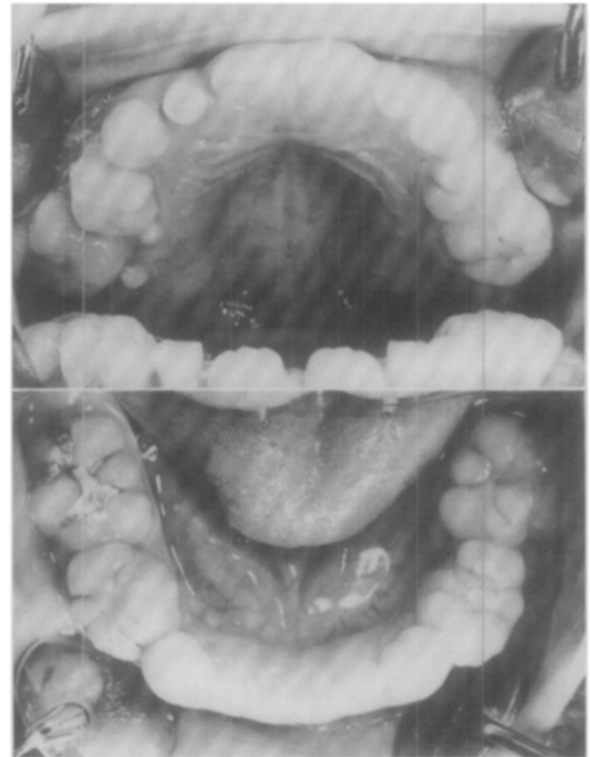


Fig. 5. Patient's dentition reveals abnormally shaped deciduous molars and two supernumerary microdontic teeth on palatal side of deciduous maxillary left molars.

Table 1. Comparison of the mean mesiodistal crown diameters of deciduous teeth in Chinese boys¹⁵ and our patient

Chinese boys (mm)	Present case	
	Left side (mm)	Right side (mm)
Maxilla		
Central incisor	6.61	7.12
Lateral incisor	5.52	5.18
Canine	6.57	7.92
First molar	7.45	9.47
Second molar	9.39	12.43
Mandible		
Central incisor	4.52	4.15
Lateral incisor	4.74	4.45
Canine	5.94	8.35
First molar	8.08	14.25
Second molar	10.26	15.48

molars had erupted. Although all posterior teeth were large and deformed, alignment was good (Fig. 5). Attrition was noted on the occlusal surfaces of the molars. Amalgam restorations had been placed in the mandibular right first and left second molars. However, all mandibular molars

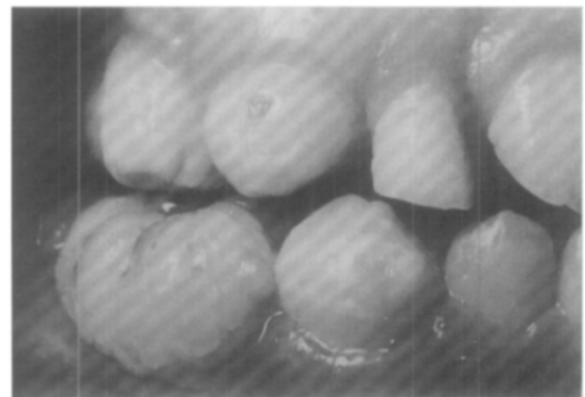


Fig. 6. Heavy dental plaque was deposited on constricted cervical area of deciduous mandibular right first molar. Also note spotlike defects on labial surfaces of canines of both jaws.

showed incipient caries in the developmental grooves. Heavy dental plaque was found on the constricted cervical area of the mandibular first molars (Fig. 6). In addition, conically shaped supernumerary microdontic teeth were present on the palatal side of each deciduous maxillary left molar (Fig. 5). Occlusion was compatible with an Angle Class III molar relationship.

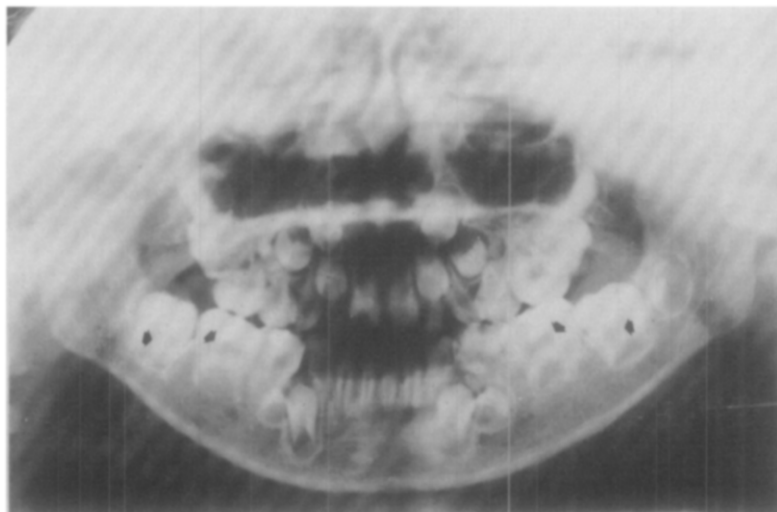


Fig. 7. Panoramic radiograph of jaws at age 7 years and 10 months shows both taurodontic configuration and vertical septum in pulp chamber of mandibular molars (*arrows*). Primordia of mandibular second premolars are still absent.

Dental radiographs showed the roots of the deciduous molars to be disproportionately short compared to their crowns (Fig. 7). Developing permanent mandibular first molars were taurodontic. A vertical septum was noted in the pulp chamber of each mandibular deciduous second and permanent first molar; thus the pulp chambers appeared duplicated. The primordia of the mandibular second premolars and the permanent mandibular right second molar still could not be seen.

Mesiodistal crown diameters of the deciduous teeth were measured with a vernier caliper (measuring to 0.05 mm) on poured casts by the method of Moorrees.¹⁴ Data were compared with the mean for unaffected Chinese boys.¹⁵ Mesiodistal crown diameters of all deciduous canines and molars of this boy were much greater than normal (Table I). However, the boy's deciduous maxillary lateral incisors and all deciduous mandibular incisors were smaller than those of unaffected Chinese boys.

His parents and a 5-year-old sister were examined; they had no morphologic dental anomalies or congenitally absent teeth. Other relatives reportedly had normal teeth and normal hearing but were not examined.

Pure-tone air-conduction audiometry was performed on the patient, his parents, and his sister in a soundproof booth. Results were normal. The father's hearing threshold, although normal at the other frequencies, dropped to 65 dB at 8000 Hz. However, he had worked in a noisy factory for many years.

DISCUSSION

Table II summarizes previous reports of otodontal dysplasia as well as our own. Autosomal dominant inheritance was demonstrated in all reported kindreds in which several individuals were affected.^{1-7, 9, 12-13} Griffin's¹⁶ patient did not have otoden-

tal dysplasia, because he did not have the typical dental abnormalities of this syndrome.

Dental anomalies and hearing loss usually co-exist in individuals with otodontal dysplasia. However, some patients may have the dental anomaly only,^{3, 5, 7, 11} because onset of hearing loss may vary from early childhood to middle age.^{3, 7} Thus it is likely that hearing loss will develop later in life in our patient. The proband of the family reported by Jorgenson and colleagues⁷ was first thought to have normal hearing but proved to have progressive hearing loss at 6¹/₂ years of age.¹⁷ Another patient who had the dental abnormalities of otodontal dysplasia was depicted with a photograph of the teeth only; hearing status was not mentioned.¹⁰

Hearing loss (hearing threshold greater than 25 B in one or both ears) in affected individuals may be found at frequencies from 500 to 3000 Hz^{3, 5, 9} or at all tested frequencies.¹²⁻¹³ Cook and coauthors¹⁷ reported that the sensorineural hearing loss may plateau by the fourth decade and result in a severe, flat, and bilaterally symmetric loss. They also proposed that the lesion was located in the cochlea but that the mechanism for developing the loss was unknown. Hearing loss in our patient's father may have been caused by the noisy environment he had worked in for many years. He had no hearing-impaired relatives.

Other dental anomalies have been noted in persons with otodontal dysplasia. Eruption of affected deciduous canines and molars is often delayed until after age 2 years,^{2, 3, 5-7} and complete eruption of the deciduous dentition may be delayed, as in the boy

Table II. Kindreds or sporadic cases of otodontal dysplasia reported by other investigators and by present authors

Family number	Investigator	Ancestry of affected patient(s)	Mode of inheritance	No. of proved affected cases	
				HL and DA	DA only
I	Levine et al. ^{1,3}	Italian	AD	26	3
II	Gundlach et al. ^{4,6}	Polish	AD	2	1
III	Jorgenson et al. ⁷	German	AD	13	1†
IV	Rapp and Winter ^{8,9}	British	AD	5	
V	Magnusson and Persliden ¹⁰	?	?	?	1
VI	Stewart et al. ¹¹	Irish	?		1
VII	Beck-Mannagetta et al. ^{12,13}	Austrian	AD	3	
VIII	Chen et al.*	Chinese	?		1

HL = coincide of hearing loss; DA = dental anomaly; AD = autosomal dominant.

*Present authors.

†The proband was proved to have hearing loss subsequent to the original report.¹⁷

reported here, until age 6 years.^{9,12} Eruption of the permanent posterior teeth may also be delayed.^{2,9,12} Winter⁹ reported a woman with the syndrome in whom the mandibular permanent first molar did not erupt until age 29 years.

Canines and molars have been reported to be deformed and large in this syndrome.^{2,3,5-13} We confirmed this finding by measuring the mesiodistal diameters of these teeth on our patient's dental casts (Table I). Incisors have been reported normal in shape and size but have never been previously measured. In our patient mesiodistal crown diameters of the deciduous maxillary lateral incisors and the deciduous mandibular incisors were found to be smaller than those of Chinese boys¹⁵ without the syndrome.

Many previous reports of otodontal dysplasia,⁵⁻¹⁰ as well as our study, describe an enamel defect on the labial surface of the canines. Witkop and coauthors⁵ disclosed, by ground section, that the defect consisted of reduced enamel thickness, enamel hypomaturation, and irregularities of adjacent dentinal tubules. There may also be considerable variation in the shape of affected molars among patients. Some patients have molars that are dome shaped with absent or shallow fissures and cusps⁸⁻¹¹; other patients, like ours, may have molars with multiple prominent lobules.^{2,3,5-7,12,13} The reason for these differences is unknown. In addition, deformities of the deciduous posterior teeth are more severe than those of the permanent dentition.

Small or absent premolars may be the only dental abnormality in otodontal dysplasia.³ In the normal individual dental crypts of the first and second

premolars should be discernible on radiographs by ages 2 and 3 years, respectively,¹⁸ and a tentative diagnosis of aplasia of a premolar can be made by age 6 years.¹⁹ In our patient calcification of the crowns of the maxillary premolars and the mandibular first premolars was delayed until after 4 years of age. His mandibular second premolars still cannot be seen on radiographs at age 7¹⁰/₁₂ years and may be congenitally absent.

Radiographic features of the teeth in otodontal dysplasia are striking. Findings consist of large and apparently duplicate pulp chambers, pulp calcifications, and taurodontic molars.^{2,3,5} Beck-Mannagetta and associates¹² disclosed by ground section that the apparently duplicated pulp chambers each had a longitudinal dentin septum. Witkop and coauthors⁵ reported that the taurodontic molars were due to enlarged crowns and pulp chambers coronal to unaffected roots rather than apically placed pulp floors, as is seen in true taurodontism. The structure of our patient's deciduous molars is compatible with this hypothesis, but the configuration of his developing permanent mandibular first molars fulfills the criteria of true taurodontism.²⁰

Other anomalies that may be associated with otodontal dysplasia are congenital coloboma of the eye,⁹ complex odontoma,^{12,13} and supernumerary microdontic teeth.⁹ Our patient had supernumerary microdontic teeth.

Histologic examination of abnormal teeth in otodontal dysplasia has revealed aberrant morphodifferentiation but normal histodifferentiation.^{5,12} Levin and coworkers³ have suggested that a defect in the dental papilla could account for the multiple con-

comitant dental anomalies. However, the etiology of otodontal dysplasia remains unknown.

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