

## Odontodysplasia in a pediatric patient: literature review and case report

Rolando Guzman, DDS, MDS Margaret A. Elliott, DDS, MPH  
Karen M. Rossie, DDS, MS

### Abstract

*A case of a three-year-old Caucasian male with odontodysplasia is presented. Although the etiology of this condition is unknown, this anomaly involves both the mesodermal and ectodermal dental components and results in deficient and abnormal formation of dentin and enamel. The orofacial characteristics and dental findings of the condition are presented in this case report.*

### Review of the Literature

Odontodysplasia is a rare developmental anomaly involving both mesodermal and ectodermal dental components in a group of contiguous teeth. The condition is characterized by deficient and abnormal formation of both dentin and enamel, and it may involve both primary and permanent teeth (Gardner 1974).

Clinically, the affected teeth appear discolored, hypoplastic, hypocalcified, and tend to be smaller, with short roots, open apices, and abnormally wide pulp chambers (Gardner and Sapp 1973). Frequently, these teeth demonstrate a failure to erupt completely. Radiographically, the teeth present with poorly discernible and deficient enamel and dentin, blotchy calcification patterns, enlarged pulp chambers, and broadened peripheral radiolucent zones (Zegarelli et al. 1963). Due to the fuzzy, radiolucent appearance, the term "ghost teeth" has sometimes been applied to this condition.

Histologically, the connective tissue which corresponds to the dental sac is loose and fibrillar, with calcified bodies scattered throughout (Sapp and Gardner 1973). The enamel organ demonstrates two different zones: one presenting a normal prismatic structure, and the other a hypoplastic globular structure consisting of smaller, closely packed crystallites (Kerebel and Kerebel 1981). The dentin appears hypoplastic with clefts

and interglobular dentin in milder cases; the more severely affected teeth exhibit cellular dentin and amorphous areas within the coronal dentin (Gardner 1974).

The first report of this condition was published by McCall and Wald (1947) under the title of "Arrested Tooth Development", and their report presented only radiographic findings. Suher et al. (1953) used the same terminology to describe another case in which histological, clinical, and radiographic findings were presented. The term "shell teeth" first was used by Rushton (1954). Chaudhry et al. (1961) utilized the term "odontogenesis imperfecta" to report a case in which this type of anomaly was present in both the maxillary and mandibular arches. Bergman et al. (1963) published a detailed histological study under the title of Unilateral dental malformation. The term "odontodysplasia" first was used by Zegarelli et al. (1963), and it has become the accepted designation for this condition.

According to the accepted definition, odontodysplasia is a developmental anomaly affecting both primary and permanent dentitions. Generally, it affects only one arch, with the maxilla being affected twice as often as the mandible (Lustmann et al. 1975). The condition usually is unilateral with no tendency to cross the midline. However, in approximately 16% of the reported cases, there was evidence of odontodysplasia affecting both maxillary central incisors; three cases described damage which crossed the midline of the mandible, affecting several teeth on both sides (Lustmann et al. 1975). Odontoblastic lesions appear to be more common in the anterior segments and affect incisors and canines more often than posterior teeth. Sex distribution appears to show that females are affected more often than males (Lustmann et al. 1975).

The etiology of odontodysplasia is unknown. Several factors have been suggested by Sadeghi and Ashrafi (1981), such as:

1. local infection
2. local trauma
3. local ischemia
4. irradiation
5. metabolic and nutritional disturbances
6. vitamin deficiency
7. hyperpyrexia
8. Rh incompatibility
9. local somatic mutation
10. genetic transmission
11. local vascular defects.

In addition, Dahllöf et al. (1987) suggested that odontodysplasia may be neural in origin, and that tooth development may be associated with the ingrowth of nervous tissue.

## Case Report

A three-year-old Caucasian male was referred by his pediatrician and family dentist to the Pediatric Dentistry Department of the University of Pittsburgh School of Dental Medicine for evaluation of delayed dentition eruption in the mandibular left quadrant.

The patient was the result of a normal pregnancy and birth. Medical evaluation revealed no significant illnesses, except for 1) chronic episodes of otitis media which resulted in four myringotomy procedures, and 2) surgical correction of a double hernia. On physical examination, the patient appeared to be well developed and well nourished and was measured to be in the 20th percentile on the Stuart and Meredith (1946) growth curve. No hereditary or systemic abnormalities were reported; however, there was a familial history of mandibular supernumerary incisors on the mother's side of the family.

Oral examination of the patient revealed a slight underdevelopment of the left side of the mandible and a distinct facial asymmetry. A bluish, vascular lesion was observed on the left side of the lower lip. Intraorally, all soft tissues appeared normal. The maxillary primary teeth were completely erupted and normal in size and shape. In the mandible, the left primary canine and the first and second left primary molars appeared to be unerupted. The mandibular primary centrals and the left primary lateral incisor appeared small and malformed. Upon questioning, the mother revealed that the mandibular left first primary molar had developed an abscess and had been extracted previously. There was no history of radiation therapy, trauma, or other dental treatment in the affected left mandibular area. Oral hygiene appeared good, and no dental caries or restorations were present.

Radiographically, the teeth in the mandibular left quadrant appeared underdeveloped. They demonstrated large pulp chambers with thin layers of enamel and dentin. No visual demarcation was present between enamel and dentin in the area of the DEJ. Overall radiodensity of the affected teeth was less than that of normal teeth (Figs 1 & 2, facing page). There was no evidence of developing tooth buds for the mandibular left first and second premolars and mandibular left permanent canine. The permanent tooth buds of the mandibular incisors were present; however, the left lateral incisor appeared delayed in development. Calcification of the mandibular left first permanent molar was delayed as compared to its contralateral molar (Fig 3, facing page).

## Treatment

A comprehensive medical and dental examination was conducted at the initial appointment, and a diagnosis of odontodysplasia was made based upon clinical presentation. Due to the concern of the mother, the patient was placed on a four-month recall schedule. Two months after the initial exam, the patient presented as an emergency with pain associated with the unerupted mandibular left second primary molar. A diffuse soft tissue swelling was present in that area. The patient was placed on antibiotics and scheduled for surgical extraction of that tooth. Due to poor patient behavior, treatment was facilitated by using Ketamine HCl for the surgical procedure. Extraction was uncomplicated and recovery was uneventful. Routine recall examination revealed no changes in intra- or extra-oral findings.

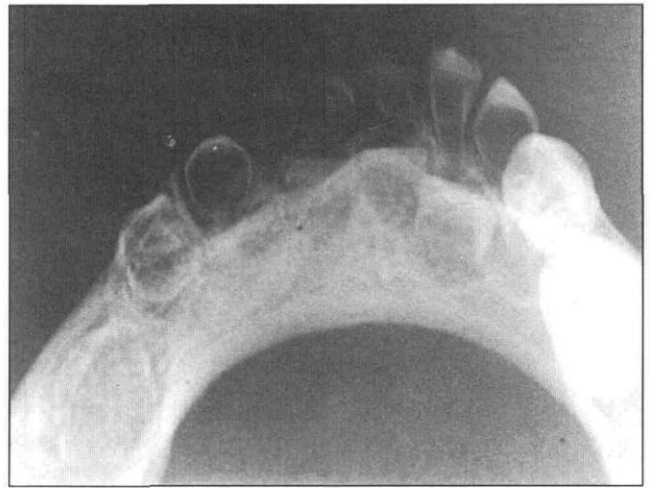
Five months after the recall appointment, the patient presented with pain and swelling in the left mandibular region. Examination revealed abscess formation associated with the mandibular left primary canine and the mandibular right central primary incisor. These teeth were extracted. During the left mandibular inferior alveolar nerve block, blood was aspirated on multiple injection attempts. Extractions were uncomplicated and recovery was uneventful.

The patient again presented with an emergency three months after the last surgical appointment. The mandibular left lateral primary incisor was fractured at the gingival margin and was painful to all stimuli. The tooth was extracted, and again, during the mandibular inferior nerve block, blood was aspirated. Extraction and recovery were without incident.

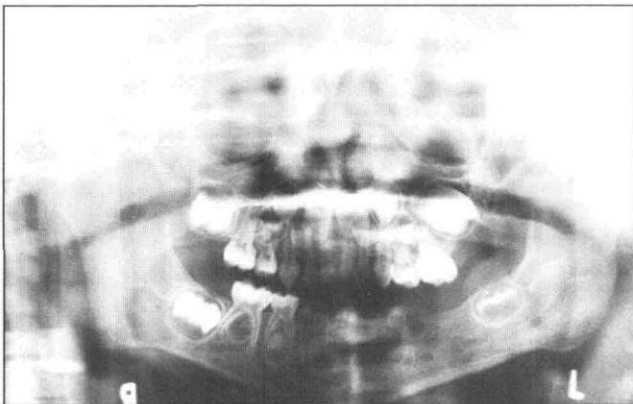
All extracted teeth were submitted for histological evaluation. On gross inspection, the teeth appeared small, hypoplastic and had underdeveloped crowns and atypical root morphology (Fig 4, facing page). On microscopic examination, the dentin appeared thinner than normal and was composed of wide, irregular dentinal



**Fig 1.** Periapical radiograph of patient at 2 years 9 months of age showing the affected mandibular left primary canine, first and second primary molars. Note the thin layers of enamel and dentin, enlarged pulp chambers, and lack of root development.



**Fig 2.** Occlusal radiograph of the mandible showing the affected primary teeth.



**Fig 3.** Panoramic radiograph of patient at 3 years 11 months of age. Note the mandibular left first and second permanent premolar tooth buds are missing, and the first permanent molar appears to be affected.



**Fig 4.** Mandibular left second primary molar affected by odontodysplasia. Note rough and irregular enamel surface, underdeveloped crown, and atypical root morphology.

tubules with areas of globular, cellular, and amorphous dentin. The predentin layer was relatively wide and exhibited proliferations of cellular dentin extending into the pulp. The pulps of these teeth were necrotic, and the pulp chambers were infiltrated with mononuclear inflammatory cells and focal collections of neutrophils. The overall histological picture was characteristic of odontodysplasia.

At the subsequent recall appointment, impressions were obtained for fabrication of a removable partial denture, and during examination, dental caries was diagnosed on the maxillary left second primary molar. During the routine administration of maxillary infiltration of Xylocaine®, the left side of the face became swollen and discolored. Ice packs were applied, and the swelling resolved in 24 hr.

A mandibular removable partial denture, replacing all affected teeth, was fabricated, and at last recall appointment the patient was wearing the appliance

intermittently. The patient was then placed on four-month recall for observation of this condition.

## Discussion

Odontodysplasia is a rare developmental anomaly that affects both the dentin and enamel of a group of contiguous teeth. The patient in this case report exhibits many of the common clinical, radiographic, and histologic features consistent with the diagnosis of odontodysplasia.

This case presents some variations from the usual clinical picture. According to Lustmann et al. (1975), teeth in the maxillary arch are affected more frequently by odontodysplasia, and this condition rarely crosses the midline. As demonstrated in this case, odontodysplasia can occur in the mandible, and teeth on both the right and left sides of the mandible can be affected. Although the literature suggests that females are af-

ected more frequently by this anomaly, it is seen in this case that males also can exhibit odontodysplasia.

An interesting clinical finding in this case involves the frequency of blood aspiration during routine injections and the episode of hematoma formation subsequent to infiltration anesthesia administration. Lustmann et al. (1975) reported a case of odontodysplasia with associated superficial hemangioma in the preauricular area and cheek. Walton et al. (1978) reported three cases of odontodysplasia with patients exhibiting vascular nevi overlying the adjacent skin of the face, and Chaudhry et al. (1961) reported a patient with birthmarks on the chin, cheek and upper neck. In the present case, a small, bluish vascular lesion was observed on the left side of the lip, and there was hematoma formation subsequent to anesthetic injection. This apparent vascular abnormality appears to be a consistent and commonly recorded finding among patients with odontodysplasia, and may suggest the consideration of vascular disturbances as a possible causal factor in the development of odontodysplasia.

Future dental care for this patient will consist of a strong preventive program and evaluation of erupting permanent teeth in the lower left mandibular area. Abscess formation and infection are a common clinical finding and may necessitate extraction of affected teeth (Neupert and Wright 1989). Due to the suspected vascular abnormality associated with this condition, special care should be taken during any surgical procedure. We will guide the occlusal development and fabricate permanent prosthetic replacements based upon the patient's overall development.

Dr. Guzman is a resident, department of orthodontics; Dr. Elliott is an associate professor, department of pediatric dentistry; and Dr. Rossie is an assistant professor, department of diagnostic services; all are at the School of Dental Medicine, University of Pittsburgh. Reprint requests should be sent to: Dr. Margaret A. Elliott, dept. of pediatric dentistry, School of Dental Medicine, Univ. of Pittsburgh, Pittsburgh, PA 15261.

- Bergman G, Lysell L, Pindborg J: Unilateral dental malformation. *Oral Surg* 16:48-60, 1963.
- Chaudhry A, Wittick H, Stickel F, Holland M: Odontogenesis imperfecta: report of a case. *Oral Surg* 14:1099-103, 1961.
- Dahllöf G, Lindskog S, Theorell K: Concomitant regional odontodysplasia and hydrocephalus. *Oral Surg* 63:354-57, 1987.
- Gardner D: The dentinal changes in regional odontodysplasia. *Oral Surg* 38:887-97, 1974.
- Gardner D, Sapp J: Regional odontodysplasia. *Oral Surg* 35:351-65, 1973.
- Kerebel B, Kerebel L: Enamel in odontodysplasia. *Oral Surg* 52:404-10, 1981.
- Lustmann J, Klein H, Ulmansky M: Odontodysplasia: report of two cases and a review of the literature. *Oral Surg* 39:781-92, 1975.
- McCall J, Wald S: *Clinical Dental Roentgenology*. Philadelphia: WB Saunders Co, 1947 pp 169-70.
- Neupert E, Wright J: Regional odontodysplasia presenting as a soft tissue swelling. *Oral Surg* 67:193-96, 1989.
- Rushton M: A new form of dentinal dysplasia, shell teeth. *Oral Surg* 7:543-49, 1954.
- Sadeghi E, Ashrafi M: Regional odontodysplasia: clinical, pathologic, and therapeutic considerations. *J Am Dent Assoc* 102:336-39, 1981.
- Sapp J, Gardner D: Regional odontodysplasia: an ultrastructural and histochemical study of the soft tissue calcifications. *Oral Surg* 36:383-92, 1973.
- Stuart H, Meredith H: Use of body measurements in the school program. *Am J Public Health* 36:1365-86, 1946.
- Suher T, Jump E, Landis R: Localized arrested tooth development. *Oral Surg* 6:1305-14, 1953.
- Walton J, Witkop C, Walker P: Report of three cases with vascular nevi overlying the adjacent skin of the face. *Oral Surg* 46:676-84, 1978.
- Zegarelli E, Kutscher A, Applebaum E, Archard H: Odontodysplasia. *Oral Surg* 16:187-93, 1963.

## Xylitol protection lasts

Children who chew gum sweetened with xylitol not only have fewer cavities than those who don't, but they continue to benefit from the gum's preventive effects for as many as two to three years, according to a recent follow-up study.

According to a University of Michigan School of Dentistry professor, children who chewed the gum three times a day and those who chewed no gum were studied over a three-year period. The children lived in a community in Finland where the basic caries prevention plan was already very good.

Overall, the gum-chewing children had 45% fewer cavities than did the control group. When the children were re-examined on follow-up at two or three years, the preventive effect remained approximately 50%.