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CASE reports

Asymmetrical tooth defects observed in hypoplastic primary teeth and amelogenesis imperfecta: case reports

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Abstract

Two rare cases of asymmetrical tooth destruction affecting the primary dentition and 1 case of hypoplastic, vertical striping X-linked amelogenesis imperfecta which affects the permanent dentition in an asymmetrical manner, are presented. The etiology of the conditions are discussed with respect to inheritance, pre-eruptive and posteruptive influences. The asymmetrical gross tooth destruction reported in the 2 primary dentitions may be multifactorial in origin but genetic influences cannot be excluded.

Variations in the number and form of teeth are, in general, genetically determined, but modifications may also arise as a result of a number of other influences. For example, the absence of teeth such as lateral incisors or third molars may be an inherited anomaly—as may other conditions such as amelogenesis imperfecta and dentinal dysplasia. On the other hand, pre-eruptive influences such as systemic illnesses or local trauma may result in alteration of the tooth bud components during tooth formation.

Dental structures generally are well protected from damaging influences during intrauterine life but children with premature or traumatic births tend to have a higher incidence of dental anomalies in the primary dentition (Rosenzweig and Sahar 1962). Kreshover et al. (1958) examined the prevalence of developmental dental abnormalities in primary teeth and concluded they were more common than previously thought. Microscopic examination of the teeth of both jaws showed a prominent incremental line extending over the incisal and occlusal portions of the deciduous anterior and posterior teeth. Rosenzweig and Sahar (1962) and Kreshover et al. (1958) found that children with hypoplastic teeth had a greater dmf rate than those without hypoplasia and suggested that hypoplastic teeth may be more susceptible to dental caries than normally calcified teeth. Factors that may disturb ameloblasts and result in their producing hypoplastic enamel include genetic causes, nutritional deficiency, maternal illnesses, birth injury, and trauma (Kreshover et al. 1958).

This paper presents 2 unusual cases of tooth destruction and 1 case of hypoplastic amelogenesis imperfecta and discusses the possible reasons for the atypical distribution of defects.

Case 1

A 4-year-old Caucasian female was referred to the pedodontic clinic for management of gross enamel erosion and severe wear of the primary teeth on the left side.

The medical history revealed no significant or relevant illnesses. The patient had not received any radiation treatment, was born full term, and was of average height and weight. The history of the pregnancy and postnatal period was uneventful. No unusual oral habits were elicited and the diet was well balanced with a moderate intake of sucrose, mainly in liquid form. The patient resided in a nonfluoridated area and had not taken fluoride supplements. As an infant the child favored sleeping on her left side.

The mother reported that the primary teeth in the left quadrant were stained, dull, and failed to erupt completely. At the time of eruption they were defective, with soft enamel which chipped easily, exposing the dentinal surface. Maintaining good oral hygiene around the affected teeth was difficult as the gingivae were red, swollen, bled easily, and were painful during brushing. The dentitions of the 2 male siblings were unaffected, with minimal caries activity. The mother was edentulous and described her own teeth as weak and thin with a tendency to crack

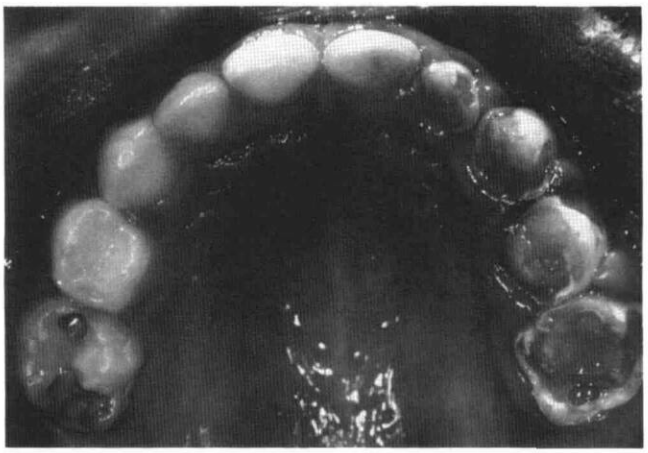

FIG 1a. Gross tooth destruction and poor gingival health was evident in the left quadrant.

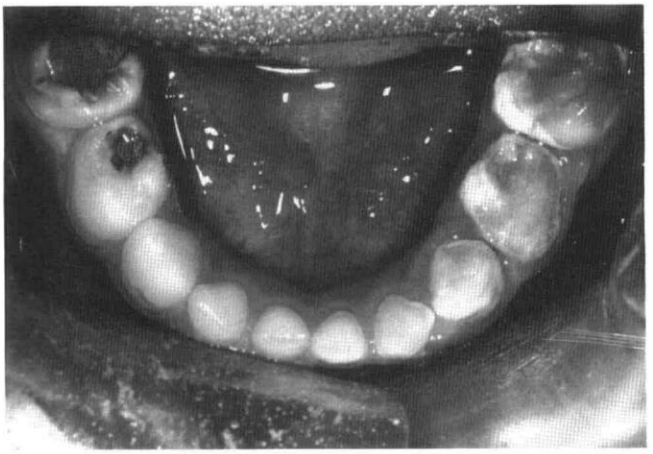

FIG 1b. Teeth in the lower left quadrant were severely decayed and gingival health was poor. The teeth in the right quadrant showed demineralization of the enamel in the gingival third.

shortly after emergence. Consequently they were removed early.

On examination the child appeared healthy, of normal height and weight with no detectable body asymmetry. The right dental quadrants seemed normal with teeth having reasonable crown height and good occlusion. The teeth in the left quadrants were severely worn, occlusal enamel was absent and exposed carious dentine was evident on the primary canines and molars (Figs 1a, b). Loss of interarch distance had occurred between the left buccal segments due to wear and reduction in crown height (Fig 2). The gingivae on the left side were inflamed, swollen, and associated with plaque deposits.

Radiographs revealed bone loss around the posterior teeth on the left side, caries, absence of maxillary second premolars, and a poorly forming upper right lateral incisor that appeared to be a peg lateral incisor (Figs 2, 3).

The treatment consisted of instructions in oral hygiene, dietary advice, systemic fluoride, and routine operative therapy. A stainless steel crown was placed on the maxillary left primary molar and after caries removal glass ionomer restorations were placed to protect the severely worn teeth. The oral hygiene improved but the gingivae on the left side remained inflamed since the patient resisted thorough cleaning in this region.

The patient was recalled regularly and by age 7 years was well motivated. Oral hygiene had improved but she developed a preference for sweet snacks. The first permanent molars emerged in all quadrants and appeared free of any defects as were the 2 lower central incisors (Figs 3a, b). The severely worn primary teeth were free from symptoms but most of the protective glass ionomer cement restorations had been worn away.

Electrophoresis of the dentine collagen peptides was performed on primary teeth on the left side of the mouth. A piece of dentine from the exfoliated lower left lateral incisor was crushed to a powder and dialysed with 24-hr changes of 0.5 M EDTA pH 7.4 at 4°C until the atomic absorption of the extracts for calcium at 422.7 nm was minimal. The residue was washed several times with distilled water, spun down, and lyophilized.

Ten mg of the freeze-dried residue was cleaved with cyanogen bromide (CNBr–substrate ratio 4:1) in 70% formic acid (substrate concentration 10 mg/ml) for 4 hr at 40°C. At the completion of the reaction the mixture was diluted with 20 Vol of distilled water and freeze dried to remove both acid and CNBr. The sample then was dissolved in 0.625 M Tris-HCl Buffer containing 2% (W/V) SDS and loaded into the sample wells of a polyacrylamide gel.

At completion of the electrophoresis the gels were stained with silver stain and then were scanned on the light table of an ultra scan laser densitometer with a fixed wavelength of 633 nm. The densitometric profiles show that the collagen obtained was identical to that obtained from normal primary dentine.

Case 2

A 2½-year-old Caucasian female was referred to the Children's Dental Clinic for management of generalized hypoplasia of the primary teeth. A medical history reported occasional ear infections which were treated with antibiotic therapy. The patient had not received any radiation therapy, was healthy, born at full term, and was of average height and weight. The child resided in a nonfluoridated area and had not taken any fluoride supplements. The mother breast-

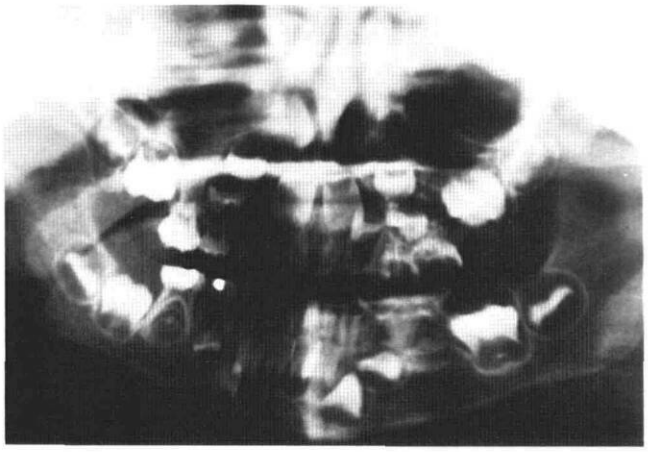

FIG 2. OPG radiograph showing absence of upper second molars and a poorly forming right lateral incisor.

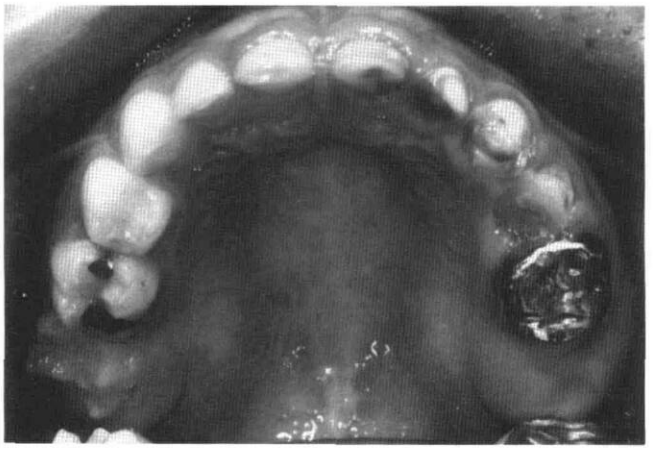

FIG 3a. Upper quadrants 3 years after the initial appointment. The first permanent molars are erupting and are free of defects.

fed her daughter until the age of 14 months, permitting the child to fall asleep during feeding. No particular side was favored during sleep. Sucrose was ingested, mainly in the form of fruit juices (they were slowly withdrawn after the initial dental appointment in the pedodontic clinic). On examination, most primary teeth were present, the upper left primary second molar was erupted and the lower left primary second molar was erupting. Minimal plaque was present and the gingivae appeared healthy. Smooth surface, early carious lesions were evident on the buccal surfaces of most teeth and the teeth on the left side were abraded. Preventive treatment was instigated and the mother was instructed in brushing the child's teeth and given dietary advice. The patient was recalled at three monthly intervals. The dentitions of two older brothers were unaffected, with minimal caries activity and no other members of the family appeared to be similarly affected.

At age 3½ years the teeth on the left side showed increased wear, while teeth on the right side were minimally abraded (Figs 4a, b). Buccal lesions were present on all teeth, with the left side more severely affected (Figs 5a, b). While caries was not detected on the occlusal surfaces of the left molars, occlusal wear was evident with dentine exposed on the first molars. The enamel covering the occlusal surfaces of the second molars on the left side was flecked and worn (Fig 4b). It appeared that the left primary second molars erupted with a defect in the enamel which was not due to poor oral hygiene or poor diet.

Case 3

A 17-year-old Caucasian female was referred to the clinic for treatment of hypoplastic permanent teeth. The condition was diagnosed as hypoplastic, vertical striping X-linked amelogenesis imperfecta. An asymmetry in the distribution of this genetic defect was observed (Fig 6). This patient also has a congenitally missing maxillary right lateral incisor. The medical history was not significant.

Discussion

Asymmetrical gross tooth destruction is unusual and may be caused by a variety of conditions. In the patients in this report, the structure of the teeth may have been affected at various stages due to genetic influences, pre-eruptive, or posteruptive factors which resulted in the asymmetrical distribution of tooth destruction.

As would be expected with a genetic disorder, the condition should affect all quadrants equally. The first 2 cases show severe tooth destruction unilaterally

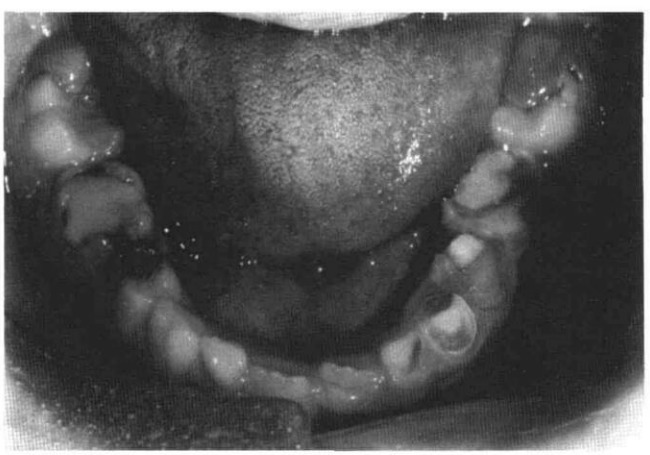

FIG 3b. The first permanent molars and central incisors are erupting in the lower quadrants. The permanent teeth appear to be free of any defect.

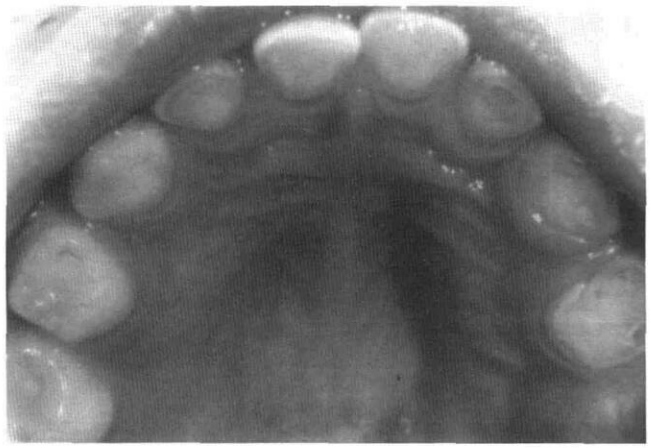

FIG 4a. All primary teeth were present in the upper quadrant. Wear of occlusal and lingual surfaces of anterior teeth was evident on the left side.

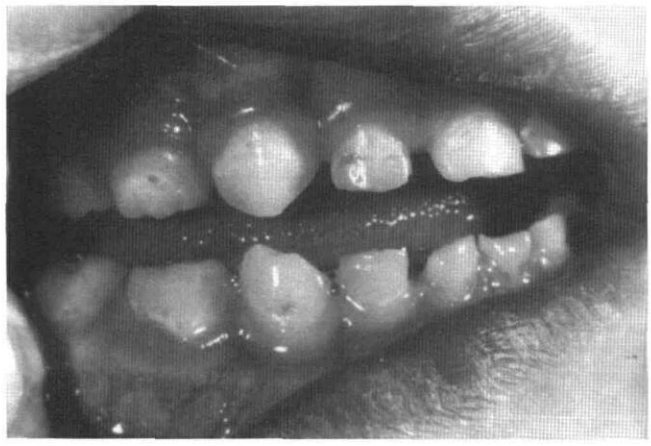

FIG 5a. Buccal lesions were present on all teeth on the right side.

and therefore, it is assumed, may exclude inheritance as a causative factor. In Case 3 it is interesting to observe that a genetic condition, amelogenesis imperfecta, affected all permanent teeth, but teeth in the right quadrants were more severely hypoplastic.

Hypodontia is often considered to be a variant of the norm, an isolated trait or an expression of a more complex syndrome (Burzynski and Escobar 1983). The genetic basis for the absence of second premolars has not been determined. However, agenesis of the maxillary lateral incisors has been reported to be an autosomal dominant trait which may be highly variable in expression and may show reduced penetrance (Shapiro and Jorgenson 1983). Variations reported include unilateral or bilateral agenesis and reduction in tooth size resulting in bilateral or unilateral peg-shaped lateral incisors.[1] Hypodontia has also been regarded as part of an X-linked disorder or as part of various syndromes[2] as observed in Case 3. Environmental factors may further complicate determination of a genetic basis for anomalies of the dentition as they may produce a phenotype similar to that caused genetically. In the first case, the absence of the maxillary premolars, the presence of a peg-shaped lateral incisor, and the mother's history of poorly formed teeth point to a possible genetic etiology despite the asymmetrical distribution of tooth destruction.

If enamel hypoplasia, due to an environmental disturbance of the ameloblasts, is a factor in the first 2 cases of tooth destruction, it may have been a local disturbance due to localized trauma rather than systemic in origin. If hypoplasia, though microscopic in

[1] Burzynski and Escobar 1983; Dixon and Stewart 1976; Shapiro and Jorgenson 1983.

[2] Brown 1983; Burzynski and Escobar 1983; Shapiro and Jorgenson 1983.

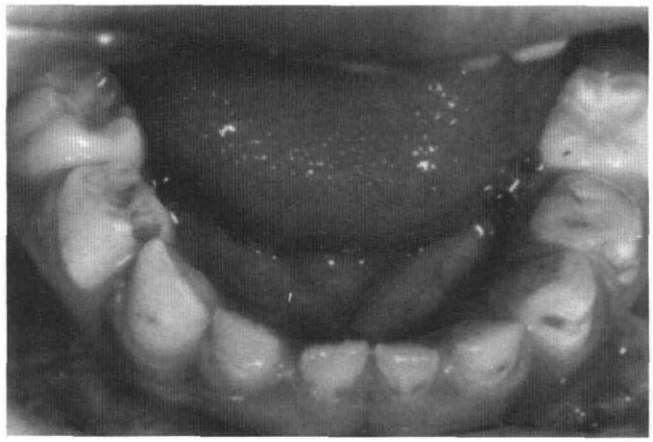

FIG 4b. Lower quadrant showing greater wear of the dentition on the left side. White flecked enamel was observed on the lower left primary second molar.

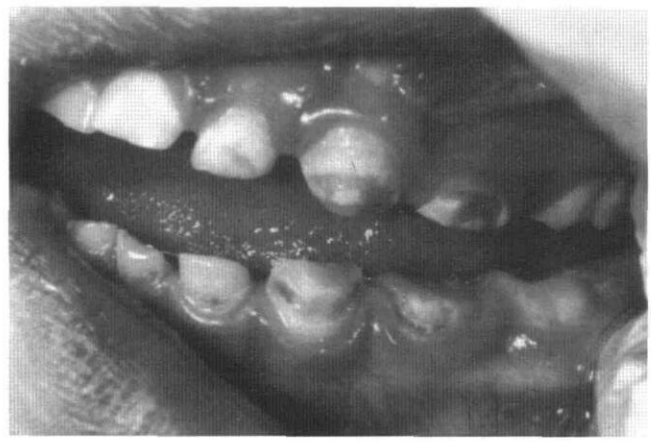

FIG 5b. Buccal lesions were more severe and affected all teeth on the left side.

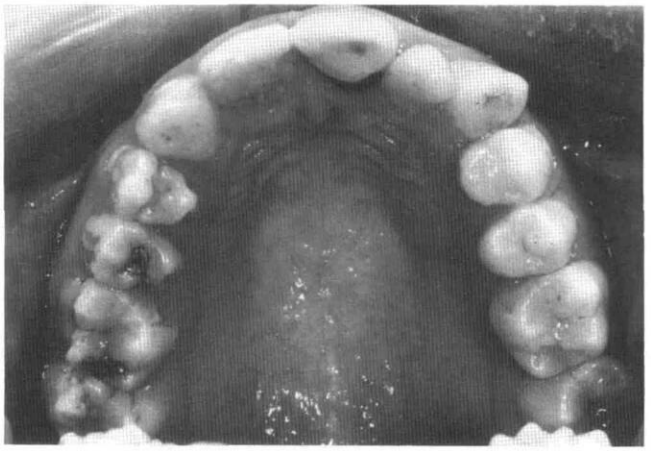

FIG 6. Hypoplasia was more severe on the right side in this patient with amelogenesis imperfecta. The right lateral incisor is congenitally missing.

size, is generally distributed to all the primary teeth, local oral habits may have increased the susceptibility of the left quadrants to caries and subsequent tooth destruction.

In nursing bottle caries the 4 maxillary anterior primary teeth are most affected while the 4 mandibular anterior teeth may exhibit no involvement at all. The canines and first molars may also be affected (Dilley et al. 1980; Ripa 1978). Drinkard and Dilley (1982) in a case report were able to describe how a child with an unusual habit of retaining a piece of banana in the mouth day and night obliterated the typical pattern of nursing caries.

In the first case of asymmetrical tooth destruction presented in this paper the patient may have favored eating on 1 side more than the other. If a pacifier was administered at bedtime, the child, lying on her left side, may have pooled food on the left side of the mouth with the teat positioned between the occlusal surfaces of the posterior teeth. The bulk of the tongue may have been pushed to the right, protecting the teeth on the right side. The mother stated that on emergence the teeth failed to form properly and chipped away as they emerged. Other parents whose children have suffered from nursing caries have made similar observations (Johnsen 1982; Kotlow 1977).

With the progression of the carious process, sensitive dentine was exposed and this possibly resulted in reduced plaque removal. Plaque accumulation and irritation on the left side would accelerate the carious process and gingival inflammation, increasing the severity of the condition on that side.

In the second case reported the lower incisors were affected by caries. It is unlikely that this caries resulted from nursing habits and may be due to caries superimposed on enamel hypoplasia of the primary dentition. The wear of the teeth on the left side may have resulted from abnormal jaw function. The greater tooth destruction on this side may have arisen from eating habits combined with reduced plaque removal.

Conclusion

The etiology of this asymmetrical gross tooth destruction may have a multifactorial basis. Although caries distribution was asymmetrical, inheritance as a causative factor for these cases cannot be excluded. In the first case the mother had apparently suffered from a form of defective tooth structure. The absence of maxillary second premolars and presence of a peg lateral incisor indicated the possibility of a genetic anomaly. Pre-eruptive influences may have resulted in hypoplasia of the enamel surface, increasing the susceptibility of the tooth structure to decalcification. Abnormal feeding habits and poor oral hygiene within the affected area may have reduced the resistance of the tooth structure to dental caries. The first permanent molar teeth appeared normal and may be maintained in good health if the patient pays special attention to oral hygiene and adopts a low cariogenic diet.

The etiology of the hypoplasia observed in the second case has not been determined but it may have resulted from a systemic illness or be of a genetic origin in which 1 side is more affected than the other.

The etiology of the hypoplasia affecting the permanent teeth in the third case is genetic. The condition is inherited as an X-linked trait with females exhibiting alternating bands of hypoplastic and normal enamel consistent with the lyonization effect of genes on the X-chromosome in heterozygous females. There is usually no homology for similarly affected teeth on each side of the mouth. The apparent increased severity on 1 side of the mouth could be due to variation in gene penetrance.

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Forensics identifies Custer's last scout

Using forensic techniques, archaeologists determined that bones and teeth found at the site of Custer's last stand in Montana were those of a mixed-blood person who was between 35 and 40 years of age, and who smoked a pipe. Michel "Mitch" Boyer, who had a French father and a Sioux mother, was the only person in Lt. Col. Custer's command who fits that description. Boyer was the cavalry leader's scout and interpreter. To substantiate the identification, archaeologists used television cameras to superimpose a picture of the bones onto the only known photo of Boyer.

The upper jaw bone was found by a tourist and more bone fragments were found when the Little Bighorn site was excavated in 1984 as part of a battlefield survey. A bullet also was found, along with buttons from civilian clothes, which the nonuniformed Boyer would have been wearing.

U.S. cigarette consumption declines

The United States Department of Agriculture reported the Americans' total cigarette consumption rose 70% from 1950 to 1981 and then fell about 9% from 1981 to 1986. The report predicts that total cigarette smoking is expected to decline the remainder of this decade because of tax increases, health concerns, and smoking restrictions.

One example of new restrictions is the government's ban of smoking cigarettes, pipes, and cigars in federal buildings, except in designated areas, which went into effect February, 1987.

Cigar and pipe tobacco use also has dropped steadily since 1970.

Use of snuff and chewing tobacco, although gaining in popularity the past decade, may decline in the rest of this decade, the report adds, because federal excise taxes have been placed on smokeless tobacco products. Also, television and radio advertisements for these products are now prohibited, and effective February, 1987, 3 rotating warning labels will appear on smokeless tobacco containers and in print advertisements.