Dental Complications of Rickets in Early Childhood: Case Report on 2 Young Girls

Vitamin D is an essential hormone for calcium gut absorption. It is also involved in child growth, cancer prevention, immune system responses, and tooth formation. Due to inadequate vitamin D intake and/or decreased sunlight exposure, vitamin D deficiency has resurfaced in developed countries despite known inexpensive and effective preventive methods. Vitamin D deficiency is a common cause of rickets, a condition that affects bone development in children and that can have serious dental complications. Deficiency during pregnancy can cause enamel hypoplasia of primary teeth. Enamel regeneration is currently impossible; hypoplasia is therefore irreversible, and once affected, teeth are prone to fast caries development. Deficiency during early childhood can affect permanent teeth and ensuing caries can sometimes lead to tooth loss at a young age. Oral manifestations of rickets should be diagnosed early by both physicians and dentists to prevent severe dental complications. This case study presents 2 young girls with rickets in early childhood who suffered from subsequent serious tooth decay. Pediatrics 2014;133:e1077–e1081
Vitamin D is an essential prohormone. Once activated by successive hydroxylations in the liver and in the kidney, it binds to the nuclear vitamin D receptor (VDR) and triggers pathways regulating calcium homeostasis, cell proliferation, and cell differentiation.1–4 Cost-effective preventive methods once eradicated vitamin D deficiency in developed countries, but it has now resurfaced worldwide and has become a major public health issue. Low, moderate, and extreme deficiencies are defined as 25-hydroxyvitamin D levels <50, <25, and <12.5 nmol/L, respectively. Moderate and extreme deficiencies result in impaired bone mineralization and ossification, leading to bone-softening diseases such as rickets, osteomalacia, and osteoporosis.1–3,5 Several types of rickets have been described on the basis of etiology, namely nutritional rickets, vitamin D–dependent rickets (VDDR), vitamin D–resistant rickets (VDDR), and hypophosphatemic rickets (HR).1 General signs of vitamin D deficiency are well known and are easy to detect on physical examination. Hypotonia, hypocalcemia, bone deformations, and lethargy are the most common symptoms.5 Vitamin D is also required for tooth mineralization. Its deficiency during gestation affects primary teeth, whereas during early childhood it affects permanent teeth. Tooth defects (enamel and/or dentin) depend on the type of rickets. Dental anomalies caused by HR have been well described.6–10 However, little is known about the dental manifestations of VDDR, VDDR, or nutritional rickets.11–14 General manifestations make early diagnosis of rickets possible. Physicians should pay particular attention to the 2 highest-risk groups: pregnant women and young children (ie, when crown formation of both primary and permanent teeth occurs). They should refer diagnosed patients to a dentist to prevent major dental complications.

Here we report the oral manifestations of nutritional rickets in 2 young dark-skinned adolescents, both of whom were treated by undergraduate and postgraduate students at the Department of Pediatric Dentistry, Charles Foix Hospital, Paris Descartes University, France. The differential diagnoses with other enamel hypoplasia etiologies are also discussed.

FIRST CASE

A 12-year-old black girl presented to the Pediatric Dentistry Department with posterior upper right jaw pain that prevented her from sleeping. Clinical and radiographic examinations revealed a deep carious lesion on her permanent upper right first molar. Emergency pulpotomy was performed after irreversible pulpitis was diagnosed. Initial medical history questioning revealed that the girl has been exclusively breastfed from birth to 7 months old, age at which fluoride and vitamin D supplements were introduced. Global oral health assessment showed deep caries on all other 3 permanent first molars as well as round-shaped enamel hypoplasia on the coronal side of both upper and lower incisors (Fig 1 A–C). Because radiographic examination confirmed the presence of all permanent teeth including third molars (Fig 1D), we decided to extract all permanent first molars and implement orthodontic treatment to correct malpositioned teeth.

SECOND CASE

An 11-year-old black girl from Cameroon was referred to the clinic by her dentist on suspicion of amelogenesis imperfecta. No similar concerns had been raised for other family members. Her health record was illegible, but her father stated that she had been treated for nutritional vitamin D deficiency with vitamin D supplements (cholecalciferol, vitamin D₃) since the age of 4 years. Her general development was otherwise normal. Clinical examination revealed severe enamel hypoplasia of incisors, canines, and permanent first molars. Premolars seemed to be intact or only slightly affected, whereas the primary lower right second molar (her single remaining primary tooth) presented enamel defects similar to those on the permanent first molars (Fig 2 A–C). Panoramic radiographic examination showed healthy lower second premolars and permanent second and third molars (Fig 2D). Anterior teeth were restored with dental composite materials, and permanent first molars were extracted.

DISCUSSION

Unlike its general complications, little is known about dental complications of rickets. Crown mineralization of primary teeth occurs from gestation to 12 months of age, as indicated in Table 1, and that of permanent teeth extends from birth to 8 years of age, except for third molars as shown in Table 2. The chronological onset of crown mineralization is tooth-group dependent. Consequently, systemic diseases or intoxications at a given period can affect various teeth differently. For example, if a chronic disease occurs in newborns, the end-stage primary tooth crown formation and the initial mineralization of permanent first molars and incisors are affected. Likewise, a similar incidence at age 3 years disrupts the mineralization of the cervical third of permanent first molars, incisors, and canines; the coronal half of first premolars; and the coronal third of second premolars. Permanent second molars may be spared. Note that these data are based on normal gestational age and are subject to slight individual variations.
FIGURE 1
A, Intraoral photograph showing slight defects on upper and lower incisors (arrows). B, Upper jaw. Note the severe decay on the first permanent molars and the presence of the second left primary molar. C, Lower jaw. Note the severe decay on the first permanent molars and the eruption of the second permanent right molar. D, Panoramic radiograph showing deep lesions on the first permanent molars.

FIGURE 2
A, Intraoral photograph showing severe defects on upper and lower incisors and canines and slight defects on the first upper bicuspid (arrows). B, Upper jaw. Note the severe enamel hypoplasia on the first permanent molars, whereas the bicuspsids are only lightly (first bicuspid) or not (second bicuspid) affected. C, Lower jaw. Note that the second primary right molar is as severely affected as the first permanent molars. Only the bicuspsids appear to be healthy. D, Panoramic radiograph showing healthy second and third permanent molars.
The first patient was not provided with vitamin D supplements from birth to 7 months of age, when crown mineralization of permanent first molars and central and lateral incisors begins. In the anterior region, only incisors displayed half-moon-shaped enamel defects. Hypoplasia affected only a reduced surface, and decay did not extend to smooth surfaces. Because the extent of dental defects is correlated with vitamin D deficiency, one can deduce that deficiency was minimal. In the posterior region, crown destruction was such that severely decayed teeth required extraction. Anomalies on the occlusal surfaces of molars make them caries sensitive, and early interception of the caries process could have therefore prevented the extractions.

The second patient suffered from more severe vitamin D deficiency. Indeed, rickets occurred throughout the first 4 years, and cholecalciferol supplementation only began at age 4 years. Tooth lesions were more extensive and mostly affected the whole crown surface of permanent central incisors and first molars. Moreover, half of the crowns of the upper lateral incisors and canines and cusps of first premolars were also affected. Given that the crown of the primary second molars mineralizes from the sixth month in utero to 10 to 12 months after birth, this deficiency may have been already present during gestation. We were unable to assess exactly when the deficiency began because the other primary teeth had already exfoliated.

Dental manifestations of nutritional rickets must be differentiated from those of hereditary VDDR, VDRR, and HR. There are 2 types (types 1A and 1B) of VDDR that are caused by mutations in the \textit{CYP27B1} and \textit{CYP2R1} genes, respectively. Both affect vitamin D-25 hydroxylase. Oral manifestations described by Zambrano et al in a 10-year-old patient were yellowish to brownish, markedly hypoplastic enamel; large pulp chambers; and short roots in all permanent teeth, as well as periodontal disease.

There are also 2 types (types 2A and 2B) of VDRR. Their etiologies are different: type 2A results from mutations in the VDR, whereas type 2B results from a normal VDR but abnormal proteins inferring with VDR ligands. Both present similar general phenotypes. Two studies have described dental anomalies in humans, that is, abnormal pulp chambers and thin dentin, as did Descroix et al in VDR-ablated mice.

HR encompasses various hereditary defects, including the following: an autosomal-dominant form (ADHR) caused by mutation in the \textit{FGF23} gene; autosomal-recessive forms (ARHR1 and ARHR2) caused by mutations in the \textit{DMP1} and \textit{ENPP1} genes, respectively; an X-linked recessive form caused by mutation in the \textit{CLCN7} gene; and an X-linked dominant form (XLHR) due to mutation in the \textit{PHEX} gene. The oral impact of the last form has been well described: enlarged pulp chambers, prominent horns extending to the dentin-enamel junction, thinner enamel, and spontaneous infectious abscesses.

Differential diagnosis of rickets with inherited anomalies such as amelogenesis imperfecta and dentinogenesis imperfecta was uncomplicated: in the present 2 cases, some teeth remained intact, whereas inherited anomalies affected all primary and permanent teeth. Other vitamin deficiencies (eg, hypovitaminosis A, C, and E) were also explored because they are all responsible for enamel hypoplasia but lead to different clinical signs. Additionally, mineral deficiencies including phosphorus and calcium deficiencies (which affect enamel and dentine formation) and magnesium deficiency (which causes enamel hypoplasia, dentine mineralization disorders, and pulp calcifications) were explored. Other possible etiologies of enamel hypoplasia, such as premature birth; celiac, renal, and congenital heart diseases; and childhood diseases such as rubella, measles, chicken pox, scarlet fever, and cytomegalovirus infections were ruled out.

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**TABLE 1** Crown Mineralization of Primary Teeth

<table>
<thead>
<tr>
<th>Tooth</th>
<th>Beginning of Crown Mineralization</th>
<th>End of Crown Mineralization</th>
</tr>
</thead>
<tbody>
<tr>
<td>Central incisor</td>
<td>3–4 intrauterine months</td>
<td>4–5 months</td>
</tr>
<tr>
<td>Lateral incisor</td>
<td>3–4 intrauterine months</td>
<td>4–5 months</td>
</tr>
<tr>
<td>Canine</td>
<td>5 intrauterine months</td>
<td>9 months</td>
</tr>
<tr>
<td>First molar</td>
<td>5 intrauterine months</td>
<td>6 months</td>
</tr>
<tr>
<td>Second molar</td>
<td>6 intrauterine months</td>
<td>10–12 months</td>
</tr>
</tbody>
</table>

Adapted from ref 15.

**TABLE 2** Crown Mineralization of Permanent Teeth

<table>
<thead>
<tr>
<th>Tooth</th>
<th>Beginning of Crown Mineralization</th>
<th>End of Crown Mineralization</th>
</tr>
</thead>
<tbody>
<tr>
<td>Central incisor</td>
<td>3–4 months</td>
<td>4–5 years</td>
</tr>
<tr>
<td>Lateral incisor</td>
<td>3–4 months</td>
<td>4–5 years</td>
</tr>
<tr>
<td>Canine</td>
<td>4–5 months</td>
<td>6–7 years</td>
</tr>
<tr>
<td>First premolar</td>
<td>1.5–2 years</td>
<td>5–6 years</td>
</tr>
<tr>
<td>Second premolar</td>
<td>2–2.5 years</td>
<td>6–7 years</td>
</tr>
<tr>
<td>First molar</td>
<td>Birth</td>
<td>2.5–3 years</td>
</tr>
<tr>
<td>Second molar</td>
<td>2.5–3 years</td>
<td>7–8 years</td>
</tr>
<tr>
<td>Third molar</td>
<td>7–10 years</td>
<td>12–16 years</td>
</tr>
</tbody>
</table>

Adapted from ref 15.
pathologies such as hypothyroidism, hypoparathyroidism, pseudohypoparathyroidism, and other systemic disorders were also screened for.17,18

**CONCLUSIONS**

Enamel defects increase the incidence of dental caries. Pediatricians, physicians, and dentists should diagnose dental manifestations of rickets early to prevent decay progression. Ideally, a first dental visit at 12 months of age enables parents to gauge the importance of oral health, feeding, and annual dental checkups.19 If parents fail to bring their children at that young age, a first visit at age 3 is essential; all primary teeth should have erupted by then. A checkup at 6 to 7 years of age allows the dentist to check for enamel hypoplasia or caries on permanent first molars and incisors. Finally, the American Academy of Pediatrics recommends vitamin D supplements for all age groups.20 Recent guidelines from the Institute of Medicine and the Endocrine Society offer additional preventive strategies.21,22

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**REFERENCES**

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