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# Primary hyperoxaluria in a pediatric dental patient: case report

## CASE REPORTS

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### Abstract

*A case is presented in which primary hyperoxaluria and oxalosis in a 14-year-old Caucasian female were diagnosed. Generalized root resorption resulted in a remarkable mobility of her maxillary central and lateral incisors, although no bone loss was noted. The management of the patient's dental concerns in this rare heritable metabolic disorder consisted of removing the maxillary incisor teeth and placing two sequential prostheses, which the patient tolerated well. A history of trauma to the maxillary incisors was ruled out, so this case adds previously unreported information to our knowledge about the effect of oxaluria on teeth and oral tissues. (Pediatr Dent 14:260-62, 1992)*

### Introduction and Literature Review

Primary hyperoxaluria, a rare autosomal recessive disorder of glyoxalate metabolism, most likely enzymatic, manifests as an excessive calcium oxalate monohydrate biosynthesis and deposition within the mesodermal tissue of the body.<sup>1-3</sup> The disorder is characterized by nephrolithiasis, nephrocalcinosis, and renal oxalate deposits. Even though nephrocalcinosis is the predominant and early clinical manifestation, extrarenal deposits of calcium oxalate monohydrate in various, but apparently selective target tissues, are a common persistent finding. These tissues may include, but are not restricted to, the kidneys, heart, ocular tissues, thyroid, bone, lymph nodes, walls of arteries and veins, brain, dentin, dental pulps, and salivary glands.<sup>4</sup> When extrarenal deposits of calcium oxalate are noted, the disorder is labelled as oxalosis. Calcium oxalate may have less of a tendency to form deposits in other tissues, but other tissues are not necessarily immune.

As the disease progresses, partial to complete loss of renal function due to nephrolithiasis is encountered, making renal dialysis necessary. Kidney transplantation is attempted frequently with varying degrees of success. The weight-bearing areas and joint spaces become affected as the oxalate accumulates in the synovial tissues and fluid of the joints, often immobilizing the patient. Anti-inflammatory and analgesic drugs are administered for pain control.

Dental and periodontal findings associated with primary hyperoxaluria and oxalosis include extensive infiltration of crystals in the pulps of developing teeth, in the marrow spaces of the alveolar bone, in the gingival corium, and in the periodontal ligament.<sup>5,6</sup> Crystalline calcium oxalate deposits within the periodontal ligament provoke a granulomatous foreign-body reaction which results in external root resorption, pulp exposure, and tooth mobility.<sup>7</sup>

### Case Report

A 14-year-old Caucasian female, diagnosed at 7 years of age with end-stage renal disease and oxalosis, presented to the Rainbow Babies and Children's Pediatric Dental Clinic with a chief complaint of painful, mobile maxillary anterior teeth with no history of trauma. The patient had received a light-cured acrylic composite splint from the maxillary right lateral incisor to the left lateral incisor from a local practitioner approximately six weeks earlier, but then was referred for definitive treatment after the splint failed as a result of insufficient enamel etching and/or excessive mobility of the incisors.

Hyperoxaluria was diagnosed based on a significant 24-hr oxalate excretion (159 mg/24 hr). There was no family history of hyperoxaluria. The patient received hemodialysis three times weekly while awaiting a renal transplant. Daily medications included 3000 units of heparin to maintain the patency of the Scribner Shunt placed in the left ankle, 0.5 mg/kg cyclosporin, 10 mg prednisone every other day, 50 mg hydrochlorothiazide, 400 mg Tagamet® (Smith Kline and French Laboratory Co., Carolina, Puerto Rico), 1.0 mg folic acid, 100 mg Pyridoxine® hydrochloride (Forest Pharmaceuticals, Inc., St. Louis, MO), 810 mg Uro-mag® (Blaine Co., Inc., Fort Mitchell, KY) in divided doses, and 250 mg Neutrophos® (Willen Drug Co., Baltimore, MD). The patient was nonambulatory and short, with yellowish brown papillary-like lesions scattered on the skin due to subdermal oxalate crystal deposits. The patient was hepatitis B-positive, but HIV negative.

Radiographs exposed at the time of presentation revealed unremarkable bony architecture, generalized loss of lamina dura, and extensive external root resorption of the maxillary central and lateral incisors (Fig 1, next page). A lateral periapical film showed a marked procumbency of the maxillary incisors, and a panoramic



**Fig 1.** Periapical radiograph demonstrates extensive resorption of the roots of the maxillary permanent central incisors. External root resorption of the maxillary lateral incisors also is evident.

film yielded no additional unusual findings. No bone resorption or bone loss was noted elsewhere in the dentition.

Both maxillary central incisors demonstrated marked mobility and severe discoloration, while the maxillary lateral incisors were of normal color and only slightly mobile. None of the maxillary incisors were sensitive to percus-

sion, but all exhibited some sensitivity to masticatory forces. Vitality testing of the incisors was not performed, because a periodontal consultation in conjunction with existing radiographs determined the teeth to be nonsalvageable. All other oral tissues appeared normal and healthy. Consultations with the patient's primary physician and hematologist before treatment led to the decision to manage the case under general anesthesia; a bone marrow biopsy also was scheduled to be performed.

Before the surgery, an immediate partial prosthesis was fabricated to act as a surgical stent and improve patient acceptance postoperatively. The surgery was on the alternate day of hemodialysis, so that the heparin administration would not affect hemostasis. The patient received penicillin G IV before and after the procedure as prophylaxis against bacterial infection of the shunt. Prednisone administration was discontinued on the day of the surgery only, due to intravenous administration of corticosteroids as a supplement.

Maxillary central and lateral incisors were extracted, and the gingiva excised and thinned under local anesthesia for hemostasis and sent with the teeth for evaluation. Sutures were placed in an interrupted fashion, and the interim prosthesis also was placed. The postoperative course was uneventful, and the patient was discharged the same day.

The permanent prosthesis, with Adams clasps on the maxillary permanent first molars and ball clasps between the premolars, was delivered 21 days postoperatively and accepted well. Since the surgery, the patient

has had a combination kidney and liver transplant to improve her condition.

## Histological Findings

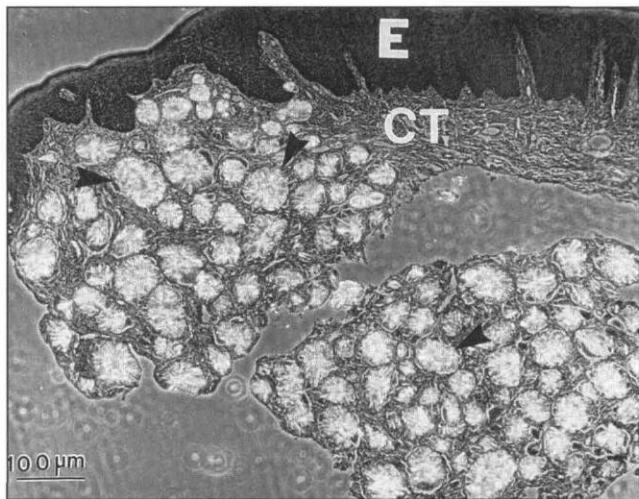
The soft tissue fragments show extensive, strongly birefringent crystalline deposits and an associated foreign body giant cell reaction (Figs 2 and 3, next page) limited to the underlying connective tissue of the gingiva. The individual deposits are roughly circular in shape, with needle-like crystals projecting from their centers, giving rise to the so-called "spokes on a wheel," and giant cells clearly are visible surrounding each crystal (Fig 3, arrows). These crystalline masses are interpreted as calcium oxalate crystals. Calcium oxalate crystals and the associated foreign body giant cell reaction were not observed within the decalcified teeth. However, the generalized root resorption and thickened predentin layer suggest the dental changes associated with oxalosis.

## Discussion

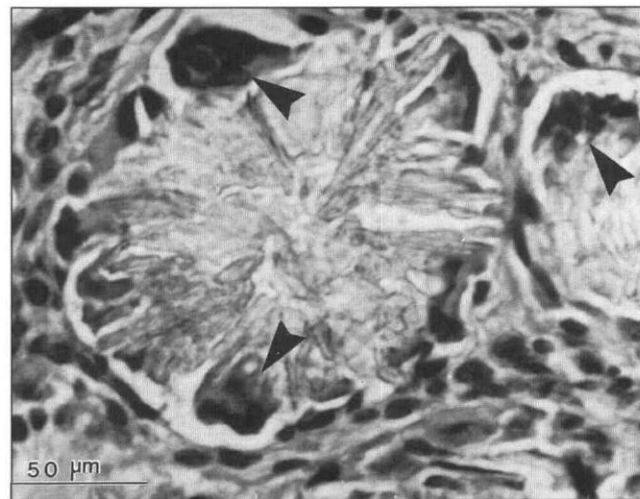
Both primary and secondary forms of hyperoxaluria are characterized by calcium oxalate deposition within the mesenchymal tissues of the affected individual. The process of oxalate deposition is not understood fully. However, Brancaccio et al.<sup>9</sup> suggested that higher plasma oxalate levels and alterations in vascular permeability may be responsible for calcium oxalate deposition. Normal pediatric values for urinary oxalate excretion are usually between 10 mg and 50 mg per 24 hr. Individuals with primary hyperoxaluria may approach a value of approximately 240 mg per 24-hr period.<sup>5, 10</sup>

The findings in this case are consistent with those of Glass<sup>5</sup> and Wysocki et al.<sup>6</sup> Fantasia et al.<sup>8</sup> offer a possible explanation for the accumulation of oxalate crystals in the periodontium.<sup>7</sup> Alteration in capillary permeability in the areas of inflammation leads to the escape of plasma proteins and oxalates into the surrounding tissues. These crystals, in turn, accelerate the inflammatory reaction (which ultimately resulted in the external root resorption seen in this condition).

Patients with primary hyperoxaluria often die early in life from renal failure caused by blockage of renal tubules.<sup>11</sup> Very little is known about this disease, because the survivor pool is so small. However, modern life-sustaining measures such as hemodialysis and peritoneal dialysis are allowing patients with primary hyperoxaluria and oxalosis to enjoy much longer and fuller lives; thus, dentists are seeing previously unencountered manifestations of this disorder. The finding of root resorption in this case report is new, but may be a sequela of the disease, since neither the child nor her parents confirmed a recent history of trauma to maxillary teeth.



**Fig 2.** Photomicrograph of a section cut from the biopsy specimen obtained from the patient's gingiva. Aggregates of birefringent calcium oxalate crystals (arrow heads) in the underlying connective tissue is evident. E: gingival epithelium; CT: Underlying connective tissue. (H & E stain; original magnification x100).



**Fig 3.** Photomicrograph of a higher magnification from Fig 1 above showing an individual rosette of calcium oxalate crystal surrounded by a connective tissue core associated with granulomatous foreign body reaction. Note the presence of giant cells at the periphery of the rosette (arrow heads). (H & E stain; original magnification x400).

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