

Gunther's Disease: Esthetic Management of Erythrodontia with 1-year Follow-up

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ABSTRACT

Intrinsic tooth stains are more permanent in nature than extrinsic stains and can be divided into two types systemic and local. Erythrodontia is reddish-brown or reddish-black discoloration of teeth caused due to congenital erythropoietic porphyria (CEP) or Gunther disease. It is the rarest type of porphyria and is usually perceived in infancy. An 8-year-old girl with erythrodontia was referred to a private clinic in the United Arab Emirates with a chief complaint of discolored primary teeth with a medical history of CEP and severe right sensorineural hearing loss. A complete esthetic rehabilitation was done.

Keywords: Congenital erythropoietic porphyria, Erythrodontia, Esthetic, Gunther disease, Maxillary anterior teeth, One-step dentin bonding system.

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BACKGROUND

The disease of CEP is an autosomal recessive form of porphyria and is frequently seen in infancy.¹ Cause of CEP is the deficiency of one of the significant enzymes in heme biosynthesis called uroporphyrinogen III synthase. CEP is usually characterized by severe scarring and blistering due to skin photosensitivity and unusually increased hair growth at the back of the hands and face.² Signs of CEP vary from mild to severe and sometimes could include extreme hair growth throughout the body (hypertrichosis), red-colored urine, reddish-black or reddish-brown discoloration of the teeth, and anemia.³ Shorter life expectancy is seen in patients with CEP due to recurring infections and hematological issues. The treatment option is a bone marrow transplant, and investigational research is still going on regarding the treatment of CEP patients with transplantation with hematopoietic stem cells and gene therapy. For the screening of CEP during pregnancies, prenatal diagnosis, and preimplantation genetic tests are available.¹

Treatment for photosensitivity is the prevention of blistering by evading light exposure that, including long-wave ultraviolet sunlight through window glass or light emitted by fluorescent sources and escaping of damage to the eyelids and cornea by wearing wrap-around sunglasses since no United States Food and Drug Administration approved treatment is available.⁴

A reddish-brown discoloration is seen in the teeth, which is caused due to porphyrin sedimented in teeth, termed erythrodontia. On exposure to long-wave ultraviolet light, teeth may fluoresce. In this case report, reddish-black discoloration of primary teeth was seen, showing erythrodontia; the patient was diagnosed with CEP having photosensitivity and anemia, so the patient had undergone bone marrow transplant at the age of 1 to increase the bone density and for chronic anemia which was associated with low vitamin D levels.⁵⁻⁷

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CASE DESCRIPTION

An 8-year-old girl reported to a private clinic with a chief complaint of discolored primary teeth with a medical history of CEP and severe right sensorineural hearing loss. The patient had undergone a bone marrow transplant at the age of 1 year for CEP.

On intraoral examination, delayed eruption of permanent teeth was seen, and reddish-black discoloration of all primary teeth and reddish-brown discoloration of 41 (Fig. 1). An orthopantomogram showed normal dentin, enamel, and pulp. The thickness of the enamel was approximately as same as that of the dentin (Fig. 2). Appropriate treatment for such cases would be esthetic



Fig. 1: Reddish-black discoloration of teeth showing erythrodonia



Fig. 3: Postoperative–direct composite veneering



Fig. 2: Orthopantomogram revealed normal dentin, enamel, and pulp and delayed eruption

rehabilitation with zirconia crowns under general anesthesia. Since the child showed a negative behavior and parents did not consent to rehabilitation under general anesthesia, direct composite veneering (Fig. 3) was done chairside in relation to 51, 52, 53, 54, 61, 62, 63, 64, 72, 73, 74, 82, 83, and 84 using Ivoclar Vivadent Tetric N-Cream™ and the parents were explained about the possible need to redo if the restoration is broken.

At 1-year follow-up erupting 11, 21, 31, 32, 41, and 42 were seen along with hypertrophic gums in the maxillary anterior region. Composite veneers in relation to 52, 53, 54, 62, 63, and 64 were intact (Fig. 4). Since the permanent teeth were in eruption stage no treatment was done for the discoloration of 11 and 21 after the complete eruption of the permanent incisors esthetic crowns are to be placed based the cooperation of the patient.

DISCUSSION

Congenital erythropoietic porphyria (CEP) is hereditary as an autosomal recessive gene. When two copies of an atypical gene for the same trait, one from each parent, are inherited, it results in recessive gene disorders. CEP is a very rare recessive genetic disorder which is caused due to mutation in the gene that codes for uroporphyrinogen III synthase, causing to sedimentation of porphyrin in different body tissues; only 13 cases have been reported in literature.^{8,9}

Intrinsic tooth stains are more permanent in nature than extrinsic stains. Erythrodonia is an intrinsic endogenous stain that is acquired during tooth development due to porphyrin deposition.^{10,11} Intrinsic discolorations of teeth can change the structure, arrangement, and density of the enamel, dentin, and pulp. Variations in the tooth complex are seen in many conditions, such as chemotherapy, where dental abnormalities,^{12,13} like



Fig. 4: A 1-year follow-up erupting 11 and 21 and hypertrophic gums in the maxillary anterior region and intact restoration in 52, 53, 62, 63, 73, and 83

enamel hypoplasia and pitting, are noted; in fluorosis, yellow to brown discoloration is seen.^{14,15} In a few metabolic disorders, like alkaptonuria,¹⁶ black discoloration of teeth, are seen. In cases of neonatal jaundice, deposition of bilirubin can cause teeth to develop discoloration during dental calcification.¹⁷ Brownish discoloration of teeth can also be caused by ingestion of tetracycline, infection, erythroblastosis fetalis and trauma, but not reddish-brown as seen in CEP.

Discolored teeth, in this case report, are esthetically restored with composite veneering, as the chief complaint was regarding the discoloration of teeth. No heavy metals, such as copper and mercury, have been used since it is not recommended for dental restorations in patients with porphyria.⁵ Nitrous oxide sedation is considered safe for CEP patients and hence attempted, but the patient was uncooperative, so it was discontinued.⁵ Since patients with CEP have photosensitivity, exposure to light while undergoing dental treatment was kept minimal.

The patient was monitored every 3 months because the patient falls under the category of high risk. During the 1-year follow-up, the permanent teeth appeared to have delayed eruption for the chronological age of the patient, and hypertrophic gingiva was seen in relation to the maxillary anterior region. Composite veneers in 52, 53, 54, 62, 63, and 64 were intact. Hypertrophic gingiva was due to improper oral hygiene, so oral hygiene instruction was reinforced.

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that is acquired during tooth development due to porphyrin deposition.⁵ Intrinsic discolorations of teeth can change the structure, composition, and thickness of the enamel, dentin, and pulp, as in chemotherapy can also cause dental abnormalities like enamel hypoplasia and pitting,⁶ and in fluorosis, dental enamel becomes stained as yellow to brown color.⁷ Other metabolic disorders, such as alkaptonuria, may also lead to black discoloration of teeth. On account of the deposition of bilirubin, the teeth can develop discoloration during dental calcification in neonatal jaundice.⁸ Brownish discoloration of teeth can also be caused by ingestion of tetracycline, infection, erythroblastosis fetalis, and trauma, but not reddish-brown as seen in CEP.

Discolored teeth, in this case report, are esthetically restored with composite veneering, as the chief complaint was regarding the discoloration of teeth. No heavy metals, such as copper and mercury, have been used since it is not recommended for dental restorations in patients with porphyria.^{9,10} Nitrous oxide sedation was used in combination with non-pharmacological behavior management during the dental treatment because the patient was apprehensive and anxious about dental treatment. Nitrous oxide sedation is considered safe for CEP patients.^{9,11} Since patients with CEP have photosensitivity, exposure to light while undergoing dental treatment was kept minimal.

The patient was monitored every 3 months because the patient falls under the category of high risk.¹² During the 1-year follow-up, the permanent teeth appeared to have delayed eruption for the chronological age if the patient and hypertrophic gingiva was seen in relation to the maxillary anterior region. Composite veneers in 52, 53, 54, 62, 63, and 64 were intact. Hypertrophic gingiva was due to improper oral hygiene, so oral hygiene instruction was reinforced.

Erythrodonia is a very unusual symptom that distinguishes CEP from other types of porphyria, such as porphyria cutanea tarda. Esthetic management of erythrodonia not only improves a child's smile to be esthetically acceptable, but also reassures the parents. Pediatric dentists can offer the most acceptable esthetic outcome in such a situation using various alternate options available, from bonding resin to esthetic crowns.

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