

Functional and Aesthetic Rehabilitation of a Patient of Enamel Renal Syndrome

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ABSTRACT

Enamel renal syndrome is a very rare disorder associating amelogenesis imperfecta with nephrocalcinosis. The characteristic feature of this syndrome is enamel agenesis and medullary nephrocalcinosis. This paper describes enamel renal syndrome in a male patient born in a non-consanguineous family with complete mouth rehabilitation.

KEYWORDS: Enamel Renal Syndrome, Amelogenesis Imperfecta, Rehabilitation

INTRODUCTION

Amelogenesis imperfecta (AI) is a term given to a group of hereditary disorders that affects both deciduous and permanent enamel. AI has frequently been reported as an isolated finding with autosomal dominant, autosomal recessive or X-linked modes of inheritance.¹ Occasionally, AI occurs as a part of syndromes like amelo-onycho-hypohidrotic syndrome, tricho-dento-osseous syndrome, Morquio syndrome, Kohlschutter syndrome, occulo-dento-osseous dysplasia, hereditary epidermolysis bullosa, nephrocalcinosis syndrome. The combination of AI with nephrocalcinosis is called as enamel-renal syndrome, which was first reported by Mac-Gibbon in 1972.² To the best of our knowledge, only 9 other cases have been described in the literature.³⁻¹² The aim of this report is to describe a patient presenting with typical features of AI and renal disease, and to highlight the important role of pediatric dentists in recognizing this uncommon syndrome.

CASE REPORT

A 7.5year old male child reported to the Unit of Pediatric dentistry, with the chief complaint of discoloured and chipped off appearance of his newly erupting teeth. He was the first child born to healthy non-consanguineous parents, with a younger female sibling. Prenatal history revealed that the child was diagnosed with atrophic right side kidney at the age of 6 months in-utero. Child was born through caesarean surgery prematurely at around 7 months of gestation and was operated for dilatation of urethral sphincter when he was 3 months old. Medical history revealed that child was having right side atrophic kidney and left side hydrocoele, he was under treatment since then in PGIMER.

General examination revealed a positive behaviour, active, apparently healthy child with normal weight and height for his age. Intraoral examination revealed early

mixed dentition stage with all four first permanent molars and maxillary & mandibular incisors affected with enamel hypoplasia [figure1 (a-e)]. Poor oral hygiene and calculus was evident on lower anteriors. Anterior open bite without any history of abnormal habit like thumb sucking or tongue thrusting. In addition, primary molars also showed chipped off appearance in all quadrants. No history of sensitivity or pain was revealed.



Figure 1: Pre-op photographs a: right lateral view, b: frontal view, c: left lateral view, d: maxillary occlusal view, e: mandibular occlusal view

How to cite this article:

Rehman F, Goyal A, Gauba K. Functional and Aesthetic Rehabilitation of a Patient of Enamel Renal Syndrome. *Int J Oral Health Med Res* 2015;2(2):53-56.

Orthopantomogram (figure 2) revealed hypocalcified enamel with normal pulp chambers suggestive of Amelogenesis imperfect associated with systemic ailment i.e. renal disease. Examination of parents, grandparents and sibling revealed normal complement of teeth. Intraoral view of the younger sister is shown in (figure 3).



Figure 2: OPG showing normal complement of deciduous & permanent teeth with enamel deficiency.



Figure 3: Sibling's intra-oral view

Complete mouth rehabilitation of child was planned. Since the child was coming from a far place, in order to reduce number of appointments an indirect approach for aesthetic rehabilitation of his anterior teeth was undertaken.

On the day of examination and diagnosis, the importance of oral hygiene was emphasized, with disclosure of plaque and brushing and flossing demonstration. In his first scheduled appointment, oral prophylaxis was performed, and all four permanent molars were restored with stainless steel crowns owing to their longevity and success in such cases.

In next appointment, slight preparation of hypoplastic teeth was done in order to give a bevelled margin for composite to bond well. Silicone impression was made, impression poured using die stone for a composite restoration. All anterior teeth and primary molars were restored to their normal anatomy using a waste expired composite restorative material, with which a template was made using biostar sheet of thickness 2mm in vacuum machine [figure 4(a,b)]. This template was used for restoring all the teeth in one appointment [figure 5(a-f)]. Completed restorations are shown in figure 6(a-e). After complete mouth rehabilitation, the patient's parents presented with a positive feedback regarding his psychosocial living. The patient has been followed for the maintenance of oral health and for the evaluation of eruption of all permanent teeth. A definitive treatment

with porcelain crowns shall be established when facial maturity has been reached.



Figure 4: Lab procedure, a: mock build up of teeth to be restored i.e. 11,21,31,32,41,42,55,65,75,85, b: vacuum formed template

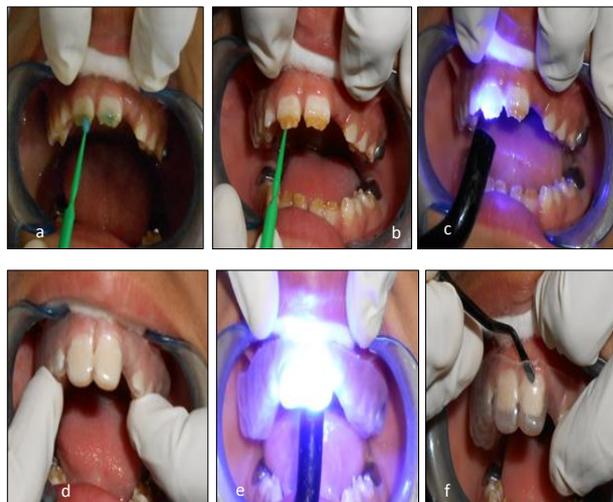


Figure 5: Clinical procedure, a: Etchant application, b: Bonding agent application, c: Polymerization of bonding agent, d: Placement of resin filled template, e: Polymerization of composite resin, f: Removal of template

Follow Up: Patient was kept on regular follow up at 3, 6, 9 and 12 months postoperatively [figure 7(a-e)]. Prognosis for the patient is good since the clinical condition was addressed at a young age prior to excessive occlusal wear.

DISCUSSION

Dental management of AI patients traditionally focuses on the restoration of the esthetics and function. However, the oral rehabilitation of children with primary or mixed dentition is more complex, since no definitive treatment can be done during periods of growth and until the eruption of permanent dentition is completely accomplished. Therefore in the present case, we have restored the anterior hypoplastic teeth with an adhesive resin which meets the aesthetic demand of the patient and posterior teeth with stainless steel crowns which suffice the functional need. Primary molars were restored in order to increase the occlusal vertical dimension and preserve the chewing function until the succeeding



Figure 6: Immediate post-op, a: Right lateral view, b: Frontal view, c: left lateral view, d: Maxillary occlusal view, e: Mandibular occlusal view

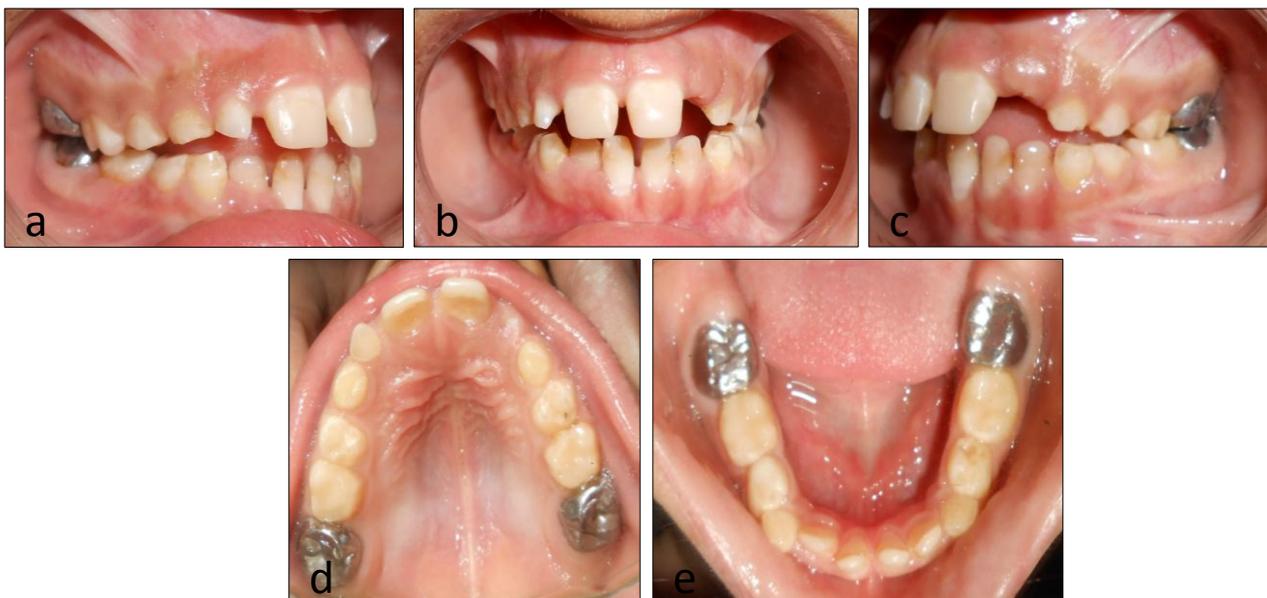


Figure 7: Follow up-6months, a: Right lateral view, b: Frontal view, c: left lateral view, d: Maxillary occlusal view, e: Mandibular occlusal view

permanent teeth erupt. Several reports have described an unusual malocclusion occurring in some AI patients that are characterized by failure of the maxillary and mandibular anterior teeth to meet in occlusion.¹² This finding was evident in the present case also. Enamel Renal Syndrome has been previously seen in 15 cases from both consanguineous and nonconsanguineous families.^{4,6-12} All these reported cases had thin or absent enamel and bilateral Nephrocalcinosis in common. While some cases progressed to renal insufficiency,^{8-11,12} others led to renal tubular disorders.^{4,6,9,11,12} Some cases

reported an association of this syndrome with other renal disorders such as polycystic kidney disease and distal renal tubular acidosis. In the present case, AI was associated with atrophic kidney which is one of its kind, being reported for the first time.

CONCLUSION

In conclusion, dentists should be aware of the presentation of Amelogenesis imperfecta associated with a systemic ailment to assist in early diagnosis and aim to provide the patient with the proper oral rehabilitation

treatment.

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Source of Support: Nil
Conflict of Interest: Nil