Amelogenesis Imperfecta -Etiology and Prosthodontic Management

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Abstract

Amelogenesis Imperfecta represents a variety of structural abnormalities of enamel that result from some malfunction of the enamel organ. The aim of the treatment is to restore esthetics, masticatory function, increase vertical dimension of occlusion and reduce hypersensitivity of teeth. Modern methods and materials have widened the range of available treatment.

KEYWORDS: Amelogenesis imperfecta, esthetics, metal ceramic crowns.

Introduction

Amelogenesis imperfecta (AI) is a heterogeneous group of hereditary disorders that affects primarily the enamel of teeth. Amelogenesis imperfecta has been described as a complex group of hereditary enamel defects that disturbs the enamel structure and exists independent of any related systemic disorder.
Although this anomaly was first described in 1890, it was not until 1938 that Finn separated it from hereditary dentinogenesis imperfecta based on clinical and radiographic evidence.[1]

CLASSIFICATION

It may be differentiated into three main groups: hypoplastic(HP) ,hypocalcified (HC) and hypomaturative (HM).
A number of classifications has been given for amelogenesis imperfecta

- **Based on clinical and radiographic features, histologic appearance and mode of inheritance:**[2]

Type 1 Hypoplastic - enamel is formed in reduced quantity but is relatively well mineralized.
Type 2 Hypo calcification -enamel is formed in normal amounts but is relatively less mineralized.
Type 3 Hypo maturation - the final stages of mineralization are abnormal

- **Classification given by Witkop (1989) [3]:**

Type	Features
I	**Hypoplastic**
IA	Hypoplastic, pitted autosomal dominant
IB	Hypoplastic, local autosomal dominant
IC	Hypoplastic, local autosomal recessive
ID	Hypoplastic, smooth autosomal dominant
IE	Hypoplastic, smooth X-linked dominant
IF	Hypoplastic, rough autosomal dominant
IG	Enamel agenesis, autosomal recessive
II	**Hypomaturation**
IIA	Hypomaturation, pigmented autosomal recessive

IIB Hypomaturation, X-linked recessive
IID Snow-capped teeth, autosomal dominant

III Hypocalcified

IIIA Autosomal dominant
IIIB Autosomal recessive

IV Hypomaturation-hypoplastic with taurodontism

IVA Hypomaturation-hypoplastic with taurodontism, autosomal dominant
IVB Hypoplastic-hypomaturation with taurodontism, autosomal dominant.

CLINICAL FEATURES AND RADIOGRAPHIC EXAMINATION

The severity of dental problems experienced by the patient varies with each type of Amelogenesis imperfecta. The primary clinical problems associated with Amelogenesis imperfecta are esthetics , teeth sensitivity and loss of vertical dimension of occlusion.

HYPO PLASTIC FORM

- In it the enamel does not develop to normal thickness.
- The crown of the teeth appear yellow brown , pitted or grooved.[4]
- Radiographic examination usually shows a full complement of teeth, but the crowns of the teeth either have very thin enamel or lack enamel completely. Enamel contrasts normally from dentin on radiographs[5].

HYPOCALCIFIED FORM

- It is characterized by poorly calcified enamel of a light yellow-brown to orange color.
- After eruption, the enamel becomes brown to black from food stains.
- The enamel of a newly erupted tooth is of normal thickness but soft and friable.
- Enamel can be easily removed from the underlying dentin.
- Enamel is less radiopaque than dentin on radiographs.

HYPO MATURATIVE FORM

- Enamel is harder with a mottled opaque white to yellow-brown or red-brown color.
- It tends to chip away from the underlying dentin.[6]
- The enamel is slightly softer than normal enamel.
- Has a mottled appearance with normal thickness at eruption.
- On radiographs its radiodensity is approximately the same as that of dentin.

ETIOLOGY

Molecular genetic studies have shown that etiology of Amelogenesis Imperfecta is related to the alteration of genes involved in the process of formation and maturation of the enamel.

Although the genetic origin of the autosomal form is less understood, the analysis of X linked Amelogenesis Imperfecta has shown the defective gene for this specific Amelogenesis Imperfecta type to be closely linked to the locus DXS85 at Xp22. Interestingly this also has been identified as the general location for human gene for amelogenin,the principal protein in developing enamel. [7]

According to Aren, some of the genes encoding the major enamel matrix proteins have been characterized; amelogenin, enamelin, ameloblastin and tuftelin. An amelogenin gene, located on the short arm of the X chromosome (Xp22.1-p22.3) has been identified for X-linked AI in some families, confirming the existence of extensive allelic heterogeneity in this condition.

Lagerstrom (1991)reported that Deletion in the Amelogenin Gene (AMG) Causes X-Linked Amelogenesis Imperfecta .He had presented the evidence that the X-linked form of this disorder is caused by a structural alteration in one of the predominant proteins in enamel, amelogenin. [8]

Collins (1999) observes the prevalence of taurodontism was similar in people with amelogenesis imperfecta and normal people. His findings suggest that some of the features associated with amelogenesis imperfecta result from abnormal enamel formation whereas others may occur as a result of expression of the genetic mutation .[9]

Wright's[10] investigation provides additional evidence that abnormal post-secretory processing of amelogenin is involved in hypomaturation and hypocalcified amelogenesis imperfecta. The unique amino acid compositions and distinct enamel protein species seen by electrophoresis and Western blot analyses suggest that different developmental processes might be involved in hypomaturation and hypocalcified amelogenesis imperfecta.

Takagi (1998) examined the size of the amelogenin peptides in the enamel of patient AI by Sodium dodecyl sulfate polyacrylamide gel electrophoresis (SDS-PAGE) and Western-blot transfer and immunobinding analysis. He suggest that amelogenin peptides constitute a major part of the protein fraction, elevating the organic content in the enamels of HCAI.

MANAGEMENT

Historically, treatment of patients with Amelogenesis imperfecta has included multiple extractions and fabrication of complete dentures.[11] The treatment plan for a patient with Amelogenesis imperfect depends upon factors such as the age of the patient, his socioeconomic status, the type and severity of the disorder and the intra-oral situation at the time of treatment.

OBJECTIVES OF TREATMENT PLAN

1. Improve the esthetics.
2. Increase vertical dimension of occlusion.
3. Restoring masticatory function.
4. Reducing hypersensitivity of the teeth.

1. Improve the esthetics

Mostly in young patients anterior permanent teeth should be treated with temporary restorations since the

pulps are large and the dentin is highly permeable and preparations for complete crowns would probably endanger the pulp.

Esthetics can be maintained with acid-etch composite resin or polycarbonate crowns.[12] The etch-retained composite resin restoration is a satisfactory alternative to the veneered metal crown. Preparation is unnecessary, and the composite resin will bond to the etched remnants of enamel.The technical process involves the selection of suitable crown forms. The teeth are cleaned, dried, etched, and primed.

Prior to treatment, the incisal edges of the lower incisors contact the lingual surfaces of the maxillary incisors, then only the labial, mesial, and distal surfaces of the crown form are filled with composite resin. In this way, premature contact will be avoided.

Composite resins, applied to dentin, have little permanent effect on the pulp.There will still be a need to protect the pulp with a calcium hydroxide preparation.[13] Shigli's (2009) [14] clinical report describes the use of composite restorations for anterior teeth for restoration of a hypomaturation type of amelogenesis imperfecta. The popularity of porcelain laminate veneers has also increased to improve esthetics.

Silicate cement offers an advantage in restoring Class III and V cavities. The range of color is extensive, and the restorations are easily replaced if the shade is not satisfactory.

Metal ceramic restorations can also be given. Metal-ceramic crowns were permanently cemented by Neelakantan on posterior and anterior teeth. Nagar P (2011) [15] treats a 24 year old patient using cast metal core in the anterior region with ceramic facing and full metal fixed partial dentures.

2. Increase vertical dimension of occlusion

Usually, treatment includes increasing the interocclusal distance by using temporary occlusal splints and employing auxiliary retention for anterior restorations. Neelakantan (2009) describe a treatment planning in hypoplastic type of amelogenesis imperfecta in a pair of twins in which vertical dimension was increased by 2 mm with the help of an occlusal splint for a period of 4 weeks in the incisor region to restore premolar and molar teeth in a favourite occlusal relationship. The preparation of anterior teeth was delayed by three weeks to assess the new vertical dimension.

Restorative treatment in patients suffering from periodontal disease complicates the therapy. In such cases it is necessary to protect the periodontal apparatus.

David (1970) described treatment for a patient with hypocalcification type of amelogenesis imperfecta.

- Temporary acrylic resin splints are made with flanges to contain the periodontal pack and to restore the lost vertical dimension.
- Due to severe loss of height of the crowns osseous surgery was indicated. Osteoectomy and osteoplasty were performed to resculpture the bony margins .
- Two weeks later, healing was sufficient to continue treatment.
- The preparations of all teeth for full crowns were completed and the splints were relined to be used as temporary restorations.

Posterior teeth with amelogenesis imperfecta may affect bone and tooth relationships. The interocclusal space may increase or remain stable.

It seems likely that for patients with amelogenesis imperfecta, if the interocclusal distance is altered by continuous eruption, no permanent changes will occur in the bony relationships.

3. Restoring masticatory function

In children Posterior teeth may be temporarily restored with stainless steel crown forms. When the child is older, the reduced pulp permits restoration of the teeth with permanent artificial crowns. In treating amelogenesis imperfecta in children , it is important to allow for mandibular and maxillary growth by using individual restorations on teeth.[16]

Cast gold crowns[17] on the permanent molars can also be given when the pulp is not to be injured by rotary instrumentation.

Metal ceramic crown or full metal crown can be given to restore masticatory function. Prithviraj (2009) describe a case report in case of hypomaturative type of amelogenesis imperfecta in which maxillary and mandibular teeth were prepared to receive metal ceramic fixed partial dentures and crowns.

Posterior teeth in the adult patient may be restored with metal veneer crowns, the preparations must extend apical to the marginal gingiva. When the length of the crowns of posterior teeth has not been reduced, increased retention may be attained by increasing the surface area of the available axial wall. This is, easily achieved by preparing vertical grooves in the dentin with a tapered fissure bur. In preparing the tooth for a partial veneer crown, which depends, to a large extent, on mesial and distal grooves for retention, additional parallel grooves will augment the retention. Pin techniques, parallel or nonparallel, give supplement retention. Nonparallel pins provide maximal retention.

In the adult patient, attrition usually reduces the length of the clinical crown. Preparation further reduce the length of the clinical crown and create a problem in retention. Siadat and Marzieh (2007)[18] determined that IPS Empress 2 with adequate strength and good bonding properties would be a good option to restore the short prepared teeth with failed crowns.

4. Reducing hypersensitivity of the teeth

Full coverage crown either metal ceramic or all ceramic can be given.

Inspite of attaining above objectives the teeth, having little enamel and no proximal contacts, generally migrate. Thus ,orthodontic therapy can be used for more favorable relationships between crowns and roots. Ozturk (2004)[19] describes interdisciplinary treatments

including orthodontics, porcelain laminate veneers, metal ceramic fixed partial dentures and direct composite restorations.

If the remaining teeth are not having sufficient occlusocervical or incisocervical height, crown lengthening[20] or osseous surgery can be performed to increase clinical height of crown. If oral cavity having the severity of attrition till only root stump are left in the mouth, endodontic treatment and overdenture can be given.

SUMMARY

The most predictable and durable esthetic option is to restore the affected teeth with complete crowns. Though this treatment option is an invasive one involving removal of substantial tooth structure, it is still more conservative than other considered alternatives, which involve extraction of remaining teeth and placement of removable prosthesis. This treatment option however, requires the patient to maintain meticulous oral hygiene.

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