# A Retrospective Study of the Prosthodontic Management of Patients with Amelogenesis Imperfecta

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***Purpose:*** This article reports on a retrospective study on patients with a diagnosis of amelogenesis imperfecta and on their prosthodontic management, oral health status, and attitudes toward their condition. ***Materials and Methods:*** The study comprised 15 patients and consisted of a clinical evaluation and questionnaire. Clinical examination included records of types of restorations and cements used, rating of restoration quality, as well as complications and periodontal variables. ***Results:*** The 15 patients had a total of 213 prosthetic restorations. The median age of the restorations was 60 months. Following the California Dental Association's system, all restorations were rated as acceptable to excellent, with one exception. During the follow-up period, four (2%) restorations had been recemented and 16 (8%) restorations had been redone, five (2%) because of porcelain fractures and 11 (5%) because of caries; two (1%) endodontic treatments were performed after prosthetic restoration. Plaque and Bleeding Indices were 28% and 21%, respectively, while pocket depths of more than 3 mm were found at 7% of all tooth surfaces. All patients judged their condition as having affected them negatively. However, after prosthodontic rehabilitation, patients experienced an improvement in self-esteem. ***Conclusion:*** Patients with severe clinical manifestations of amelogenesis imperfecta obtained extensive prosthodontic treatment at an early age. The restorations had in general performed well, and all patients were affected positively as a result. *Int J Prosthodont 2005;18:189–194.*

Amelogenesis imperfecta (AI) is a genetically determined and rare enamel mineralization defect reported by Spokes[1] in 1890 as "hereditary brown teeth." Amelogenesis imperfecta was characterized as a clinical entity in 1945, and its clinical manifestations, histologic appearance, and genetic pattern are characterized by their heterogeneity. The enamel deficiencies can be quantitative and/or qualitative, and they can vary from affecting only a few teeth with small white spots to being more general and affecting the entire dentition. The latter is manifested as yellow-brownish teeth with a soft enamel that is easily split. Generally, both the primary and secondary dentitions are affected.

Patients with AI are often esthetically affected because of tooth discoloration, often with accompanying hypersensitivity. Other associated clinical findings are reported secondary symptoms and include delayed tooth eruption or impaction, anterior open-bite occlusion, low or high caries susceptibility, predisposition to gingival inflammation, pulpal calcification, and taurodontism.[2] Various classification systems based on genetic pattern, clinical manifestations, and histologic appearance have been presented. They range from two or three main groups (namely the hypoplastic and hypomineralized or the hypoplastic, hypomaturation, and hypocalcified types) to as many as 14 subgroups.

AI is rare, with a prevalence of 0.06:1,000 in a study of 4- to 12-year-olds in the United States[3] and 0.1:1,000 in children between 6 and 18 years old in Israel.[4] A prevalence of 0.2:1,000 is reported among 3- to 19-year-olds in the western part of Sweden[5] and 1.4:1,000

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**Table 1** Individual Characteristics of Participating Amelogenesis Imperfecta (AI) Patients

| Patient | Age (y) | Gender | Type of AI* | Type of restoration | | | | |
|---|---|---|---|---|---|---|---|---|
| | | | | Gold crown | Metal-ceramic crown | Complete crown | Onlay/ inlay | Veneer |
| 1 | 18 | F | 1 | 4 | 0 | 0 | 0 | 6 |
| 2 | 20 | F | 1 | 0 | 0 | 0 | 8 | 4 |
| 3 | 23 | M | 1 | 4 | 16 | 0 | 0 | 0 |
| 4 | 23 | M | 1 | 0 | 6 | 0 | 0 | 6 |
| 5 | 23 | M | 1 | 0 | 10 | 4 | 0 | 0 |
| 6 | 24 | F | 1 | 0 | 4 | 0 | 4 | 8 |
| 7 | 26 | F | 1 | 0 | 3 | 0 | 1 | 0 |
| 8 | 27 | M | 1 | 0 | 15 | 0 | 2 | 0 |
| 9 | 29 | M | 1 | 0 | 0 | 4 | 0 | 0 |
| 10 | 37 | F | 1 | 0 | 1 | 6 | 3 | 0 |
| 11 | 20 | F | 2 | 2 | 8 | 0 | 0 | 0 |
| 12 | 14 | F | 2 | 0 | 1 | 4 | 0 | 0 |
| 13 | 19 | F | 2 | 0 | 12 | 6 | 0 | 6 |
| 14 | 22 | M | 2 | 0 | 21 | 0 | 0 | 6 |
| 15 | 28 | M | 2 | 0 | 24 | 4 | 0 | 0 |

*Type 1 = hypoplastic; type 2 = hypomineralized.

in all children between 0 and 19 years old in the northern part of Sweden.[6]

The need for prosthodontic management of this group of patients varies. Some patients only need oral hygiene instructions, whereas others need extensive dental treatment that has traditionally included conventional gold and metal-ceramic crowns cemented with zinc phosphate. In recent years, adhesive restorations have also been used. As AI is an unusual condition, few follow-up studies have been published; these are mainly case reports that present treatment modalities and outcomes.[7–12]

The primary purpose of this study was to retrospectively study the outcome of the prosthodontic management of patients with AI, as well as to obtain information on the general oral health status of these patients. The secondary purpose was to evaluate the patients' attitudes toward their condition and the prescribed prosthetic treatment.

## Materials and Methods

### *Patients*

This study involved 15 patients, 7 males and 8 females, who had been referred to the Eastman Dental Institute, Department of Prosthetic Dentistry, The Public Dental Service in Stockholm County, Sweden (Table 1). Their median age was 23 years (range 14 to 37 years). All patients had received fixed prosthetic treatment and gave consent for participation in this study. The patients were classified into two basic groups: 10 patients as hypoplastic type (AI:1; Fig 1), and 5 patients as hypomineralized type (AI:2; Fig 2), following the classification reported by Sundell and Koch.[5] In total, 403 teeth were examined, 213 with prosthetic restorations and 190 without restorations.

### *Clinical Follow-up*

Both authors performed the clinical examinations. The follow-up appointments included patient histories, clinical and radiographic examinations, clinical photography, and impressions of the dental arches. The following dichotomized measurements were noted at the follow-up examination:

- Prosthetic status (type of crown, type of cement, and time since rehabilitation).
- The quality of the prosthetic restorations was evaluated in accordance with the Quality Evaluation for Dental Care guidelines issued by the California Dental Association (CDA).[13] This included an evaluation of the surface/color, anatomic form, and marginal integrity. The restorations were classified independently by the two authors; in cases of disagreement, a joint decision was made.
- Complications of prosthetic restorations were also registered (loss of cementation and material fracture, caries, and endodontic treatment).
- Periodontal parameters (Plaque Index [PI], Bleeding Index [BI],[14] and pocket depth [PD] of more than 3 mm) were registered at the mesial, palatal, distal, and labial surface of each tooth.

To assess all possible prosthetic complications, the authors retrospectively studied the dental records dating from the first prosthetic restoration and recorded

**Fig 1a** Anterior view of a patient with amelogenesis imperfecta, hypoplastic type (with mixed dentition).

**Fig 1b** Patient shown in Fig 1a. Maxillary teeth are restored with adhesively luted porcelain veneers and crowns.

**Fig 2a** Anterior view of patient with amelogenesis imperfecta, hypomineralized type.

**Fig 2b** Patient shown in Fig 2a. Maxillary teeth are restored with all-ceramic crowns cemented with zinc phosphate. Mandibular teeth are restored with metal-ceramic crowns in the premolar and molar regions and anterior porcelain veneers.

all ensuing complications; this means that the median age of the restorations is based from the date of cementing the first crown.

### *Questionnaire*

The clinical examination was followed by a questionnaire using a visual analogue scale (VAS) to allow the patients to give their overall opinion of their handicap and the treatment received.[15] On this scale, 0 indicated complete discontent, whereas 10 was recorded as complete satisfaction.

## Results

### *Prosthodontic Status*

The 213 prosthetic restorations (Table 2) comprised 10 gold crowns, 121 metal-ceramic crowns, 28 ceramic crowns (Procera Allceram, Nobel Biocare), 18 porcelain onlays/inlays, and 36 porcelain veneers. The median age of the restorations (n = 213) was 60 months (range 12 to 240 months); patients with AI:1 had a mean of 12 restorations (range 4 to 20), whereas patients with AI:2 had a mean of 19 restorations (range 5 to 28). All complete, gold, metal-ceramic, and ceramic crowns were cemented with zinc phosphate, whereas porcelain onlays/inlays and veneers were luted with a resin cement.

### *CDA Ratings*

Regarding surface/color, 208 of the 213 restorations were evaluated as excellent according to CDA criteria, 4 were rated as satisfactory, and 1 was regarded as not acceptable. Regarding anatomic form, 196 restorations were noted as excellent, 16 as satisfactory, and 1 as not acceptable. Marginal integrity was regarded as excellent in 166 cases, satisfactory in 46 cases, and not acceptable in 1 case. In total, 212 of 213 restorations were regarded as satisfactory to excellent, the majority as excellent in all parameters. Only 1 crown was regarded as not acceptable in all parameters.

**Table 2** Prosthetic Restorations

| Type of restoration | AI:1 | AI:2 | Total No. | Median age (mo) | Age range (mo) |
|---|---|---|---|---|---|
| Gold crown | 8 | 2 | 10 | 81 | 18–195 |
| Metal-ceramic crown | 55 | 66 | 121 | 55 | 12–240 |
| Complete crown | 14 | 14 | 28 | 42 | 36–72 |
| Onlay/inlay | 18 | 0 | 18 | 42 | 12–48 |
| Veneer | 24 | 12 | 36 | 35 | 12–48 |

AI:1 = amelogenesis imperfecta type 1, hypoplastic; AI:2 = amelogenesis imperfecta type 2, hypomineralized.

**Table 3** Causes and Frequency of Complications with Zinc Phosphate–Cemented Restorations (n = 149) and Adhesively Luted Restorations (n = 54)

| Complication | AI:1 | | AI:2 | | Total | |
|---|---|---|---|---|---|---|
| | Zinc phosphate | Adhesive luting | Zinc phosphate | Adhesive luting | Zinc phosphate | Adhesive luting |
| Loss of retention | 3 | 1 | 0 | 0 | 3 (2%) | 1 (2%) |
| Fracture | 2 | 3 | 0 | 0 | 2 (1%) | 3 (6%) |
| Caries | 0 | 1 | 11 | 0 | 11 (7%) | 1 (2%) |

AI:1 = amelogenesis imperfecta type 1, hypoplastic; AI:2 = amelogenesis imperfecta type 2, hypomineralized.

**Table 4** Periodontal Parameters (% of surfaces)

| Type of AI | Plaque Index | Bleeding Index | Pocket Depth |
|---|---|---|---|
| AI:1 | 22.5 | 15.7 | 1.7 |
| AI:2 | 40.9 | 33.3 | 17.5 |
| AI:1 + 2 | 28.3 | 21.2 | 6.7 |

AI:1 = amelogenesis imperfecta type 1, hypoplastic; AI:2 = amelogenesis imperfecta type 2, hypomineralized.

**Table 5** Questionnaire (translated)*

| Question | Median | Range |
|---|---|---|
| Are you satisfied with your teeth today? | 9.0 | 5.0–10.0 |
| Have you been positively affected by the prosthetic treatment? | 9.5 | 7.5–10.0 |
| Were you satisfied with your teeth during your adolescence? | 2.0 | 0.0–4.5 |

*Ten-point visual analogue scale used for answering the questions.

### *Prosthetic Complications*

The prosthetic complications are presented in Table 3. Loss of cementation was registered in 4 restorations. Fractures were noted in 5 restorations. One ceramic crown had fractured twice. All of these restorations were remade. Complications because of caries were registered in two patients. In one of the patients, 11 complete crowns had been redone because of caries (9 metal-ceramic crowns and 2 gold crowns), and one mesial resin composite restoration had been done on a porcelain inlay in the other patient. A total of 5 endodontically treated teeth were found in the 403 (1%) teeth examined; of these, 2 (1%) were done after prosthetic rehabilitation.

### *Periodontal Parameters*

In total, 403 teeth, 213 with restoration and 190 without restoration, were examined (Table 4). The PI was 28% and the BI was 21% at all surfaces. A PD of more than 3 mm was registered in 47% of all individuals and at 7% of all teeth surfaces. The peridontal parameters generally had a higher score in the AI:2 group compared to the AI:1 group.

### *Questionnaire*

Most patients were disturbed about the discoloration of their teeth before prosthodontic treatment, but the treatment initiative resulted in a strong positive effect on patient well-being (Table 5). The questionnaire also

**Table 6** Recommended Management of Patients with Amelogenesis Imperfecta

| Age (y) | Treatment |
|---|---|
| 0–6 | Diagnosis<br>Information to parents<br>Continuous support, with oral hygiene instructions, prophylaxis and fluoride to prevent sensitivity, gingivitis, and caries<br>If needed, use glass-ionomer or resin-composite restorations, thin provisional crowns, or orthodontic bands |
| 6–12 | Treatment planning in a multidisciplinary context<br>Supervise occlusal development, note early occlusal wear and tendency for skeletal open-bite occlusal relationship<br>Information to patient and parents as to what treatment possibilities exist<br>Continuous oral hygiene and fluoride prophylaxis support<br>Maintain function with thin provisional or permanent crowns or onlays if occlusal wear is evident<br>Maintain esthetics with resin composite restorations (but avoid labial resin composite if possible in anterior teeth if these can be restored with porcelain veneers) |
| 12–20 | Definitive treatment plan based on clinical findings, radiographic evaluation, periodontal, orthodontic, and oral surgical consultations and patient desires<br>Treatment plan presented and discussed with patient and parents<br>Start prosthetic rehabilitation at a time suitable for the patient both mentally and physiologically<br>Resin-bonded porcelain therapy can be favorable, especially in anterior teeth |

asked the patients at what age they would have preferred treatment. Seven patients chose between the ages of 10 and 15; eight patients chose a later date, but none chose above the age of 25.

## Discussion

Most of the published articles about AI deal with the frequency of the disease, genetic patterns, or treatment modalities in case reports. Few studies report the prognosis of prosthodontic treatment and patients' oral health and attitude toward their condition and treatment. Before the mid-1990's, treatment implied zinc phosphate–cemented restorations. Currently, an alternative adhesive luting technique is readily available. This retrospective study was limited to a small number of patients, as AI is a rare condition.

The overall result was a good one. Of 213 restorations, 9% failed, including both recemented and remade crowns. Recorded complications were generally comparable to those recorded in a normal population.[16,17] Cheung[16] reports a failure rate of 14% on 152 single-unit crowns made in patients with normal enamel; 2% of the restorations were recemented in both the present study as well as in Cheung's.

Five porcelain fractures were all recorded in the group of ceramic restorations (n = 82), a fracture rate of 6%. As 60% of the ceramic restorations in this study were anterior restorations, this could explain the low fracture rate observed.

One of the most frequently reported causes of failure in fixed prostheses is caries lesions.[16,17] In the present study, caries was recorded in 7% of the zinc phosphate–cemented and 1% of adhesively luted restorations.

In the current study, a total endodontic treatment frequency of 1.2% compares favorably with that reported for non-AI patients with a comparable mean age[18]; a frequency of endodontic complications of between 2% and 11% is usually reported in other studies.[16–18] The reason for this low figure may be the relatively young population with favorable pulpal vascularity in our study. On the other hand, the pulp cavity is large in young adults, which might increase the risk of damage to the pulp when tooth preparation is done.

The frequency of PI in this study corresponded to reported epidemiologic data, but the BI was lower and PD was higher when compared to corresponding age groups in published studies.[18]

If the five patients with AI:2 are studied separately, higher PI, BI, and frequency of PD were present when compared to patients in group AI:1. This is comparable with Sundell's[19] results, which also showed great variation in the parameters measured both between the two AI groups as well as within each group. This reported individual variation is confirmed in the present study.

Nearly all of the patients assessed themselves as being esthetically disturbed by their pretreatment condition, in spite of the heterogeneity of the clinical manifestations. Their condition negatively affected both their relationships with other people and their self-esteem. The prosthodontic management had a positive influence on all of the patients, and they all judged the esthetic rehabilitation as the most important improvement. Many patients also reported decreased sensitivity in their teeth and judged this to be important.

In general, most patients thought their prosthetic rehabilitation should have started at least 1 year earlier than it had, and nearly half of the group wished that the rehabilitation had started before the age of 16. This suggests that planning rehabilitation with the patient at this early age is preferable, particularly as the use of resin-bonded porcelain restorations is so accessible.

The records from the patients with a diagnosis of AI:2, the hypomineralized type, showed that they are the ones in most need of prosthetic rehabilitation in the early years, often including all teeth. This presents a major challenge to the clinician and patient because of the young age of the patients, their pronounced teeth sensitivity, inflammation of the gingival tissues, poor oral hygiene, advanced damage/attrition with no remaining cuspal structure, and low height of the dental crowns.

In this complex rehabilitation, it is essential for the prosthodontist to play a key role in the multidisciplinary team supporting both patient and parents[20] (Table 6). The preventive and initial phase seeks to reduce teeth sensitivity, prevent attrition, maintain/restore masticatory function, and improve esthetic appearance. Proposed essential parameters of the rehabilitation are:

1. Early diagnosis and sustained treatment planning in a multidisciplinary context.
2. Information to patient and parents as to what treatment possibilities are, plus planning with patient and parents regarding treatment needs.
3. Sustained support, with oral hygiene instruction and fluoride prophylaxis.
4. Initiation of prosthodontic rehabilitation at a time suitable for the patient's mental and physiologic convenience. In young patients, ensure that there is no tendency toward an open-bite occlusal relationship.
5. Resin-bonded porcelain therapy can be favorable because it gives patients prolonged improved esthetics and is often less invasive; this is particularly important in young patients.

## Conclusions

A total of 15 patients with a diagnosis of AI were examined, and their prosthodontic needs, restorations, health, and attitudes toward their handicap were evaluated.

- The 213 restorations generally performed well in the context of the CDA system, and all restorations but one were regarded as acceptable to excellent. A total of 8% of the restorations were remade, and 2% were recemented.
- Patients' endodontic and periodontal status did not differ from those encountered in a normal population, but serious individual variations regarding the periodontal status were seen, especially in the AI:2 group.
- Patients with severe clinical manifestations of AI required extensive prosthetic treatment at an early age.
- Nearly half of the patients wished that the prosthodontic rehabilitation had started before the age of 16.
- All patients were positively influenced by their prosthodontic treatment.

## References


1. Spokes C. Case of faulty enamel. Br J Dent Sci 1890;33:750–752.
2. Neville BW, Damm DD, Allen CM, Bouquet JE. Abnormalities of the teeth. In: Oral and Maxilofacial Pathology. 2nd ed. Philadelphia: Saunders, 2002:49–106.
3. Witkop CJ. Heriditary defects in enamel and dentin. Acta Genet 1957;7:236–239.
4. Chosack A, Eidelman E, Wistoski I, Cohen T. Amelogenesis imperfecta among Israeli Jews and description of a new type of local hypoplastic autosomal recessive amelogenesis imperfecta. Oral Surg 1979;47:148–156.
5. Sundell S, Koch G. Hereditary amelogenesis imperfecta. Epidemiology and classification in a Swedish child population. Swed Dent J 1985;9:157–169.
6. Backman B, Holm A-K. Amelogenesis imperfecta prevalence and incidence in a northern Swedish county. Community Dent Oral Epidemiol 1986;14:43–47.
7. Lamb DJ. The treatment of amelogenesis imperfecta. J Prosthet Dent 1976;36:286–291.
8. DeSort KD. Amelogenesis imperfecta: The genetics, classification, and treatment. J Prosthet Dent 1983;49:786–792.
9. Rada RE, Hasiakos PS. Current treatment modalities in the conservative restoration of amelogenesis imperfecta: A case report. Quintessence Int 1990;21:937–942.
10. Greenfield R, Iacono V, Zove S, Baer P. Periodontal and prosthodontic treatment of amelogenesis imperfecta: A clinical report. J Prosthet Dent 1992;68:572–574.
11. Bouvier D, Duprez JP, Pirel C, Vincent B. Amelogenesis imperfecta—A prosthetic rehabilitation: A clinical report. J Prosthet Dent 1999;82:130–131.
12. Yip H-K, Smales R. Oral rehabilitaion of young adults with amelogenesis imperfecta. Int J Prosthodont 2003;16:345–349.
13. California Dental Association. Quality Evaluation for Dental Care. Guidelines for the Assessment of Clinical Quality and Professional Performance. Los Angeles: CDA, 1977.
14. Lenox JA, Kopczyk RA. A clinical system for scoring a patient's oral hygiene performance. J Am Dent Assoc 1973;86:849–852.
15. Haskisson EC. Visual analogue scale. In: Melach R (ed). Pain Measurements and Assessment. New York: Raven, 1983:33–37.
16. Cheung GSP. A preliminary investigation into the longevity and causes of failure of single unit extracoronal restorations. J Dent 1991;19:160–163.
17. Walton JN, Gardner FM, Agar JR. A survey of crown and fixed partial denture failures: Length of service and reasons for replacement. J Prosthet Dent 1986;56:416–421.
18. Hugoson A, Koch G, Bergendahl T, et al. Oral health of individuals aged 3–80 years in Jonkoping, Sweden in 1973, 1983 and 1993. Swed Dent J 1995;19:243–260.
19. Sundell S. Hereditary amelogenesis imperfecta. Swed Dent J 1986; 10:151–163.
20. Bergendal B. The role of the prosthodontists in habilitation and rehabilitation in rare disorders. Int J Prosthodont 2001;14:466–470.