



# Full-mouth adhesive rehabilitation in a case of amelogenesis imperfecta: a 5-year follow-up case report

**David Gerdolle**, DMD, MSc

Private Practice, Montreux, Switzerland

**Eric Mortier**, DMD, PhD

Senior Lecturer, Department of Conservative Dentistry and Endodontics,

Faculty of Odontology, University of Nancy, France

Private Practice, Essey-lès-Nancy, France

**Adeline Richard**, DMD

Research Fellow in MSc, Division of Cariology and Endodontics,

School of Dental Medicine, University of Geneva, Switzerland

Private Practice, Gland, Switzerland

**Francesca Vailati**, MD, DMD, MSc

Senior Lecturer, Department of Fixed Prosthodontics and Occlusion,

School of Dental Medicine, University of Geneva, Switzerland

Private Practice, Geneva, Switzerland



Correspondence to: David Gerdolle

Avenue des Alpes 29, CH-1820 Montreux. Tel: +41 21 963 0033; E-mail: davidgerdolle@gmail.com



## Abstract

Amelogenesis imperfecta (AI) is a hereditary disorder caused by mutations of genes primarily involved in the enamel formation. Several different types of AI have been identified, based on the phenotype and on the mode of inheritance. Regardless of the type, the dental treatment tends to be the same, favoring the complete removal of the compromised enamel late in the patient's life. With the new dentistry guidelines that orient clinicians towards minimal invasiveness, it should be mandatory to intercept patients affected by AI earlier, not only to protect the dentition from further degradation but also to help patients improve their self-esteem. This article examines the restorative dentistry performed on a 24-year-old Caucasian female suffering

from the hypoplastic type of AI, using only adhesive procedures. Due to the complex needs of the patient, an interdisciplinary approach was followed, involving orthodontics, periodontics, and restorative dentistry. A full-mouth adhesive rehabilitation was achieved by means of direct composite restorations, veneer/onlays and facial/palatal veneers. No elective endodontic therapy was necessary for restorative purposes. The esthetics, mechanics, and biological success were achieved and maintained. The bond to the enamel did not show signs of degradation (eg, discoloration or infiltration) even after 5 years of function. This is encouraging as it shows that adhesive techniques may be a reliable approach even in the presence of a compromised enamel layer.

*(Int J Esthet Dent 2015;10:xxx-xxx)*





**Table 1** Most common clinical findings related to amelogenesis imperfecta (AI)

compromised periodontal health
asymmetric gingival contour
hypersensitive teeth
higher risk of cavities
discolored and pitted surfaces
diminutive teeth with short clinical crown height
malformed teeth
congenitally missing teeth
pulp calcifications
taurodontism
anterior and posterior open bite
multiple posterior diastemata
loss of vertical dimension occlusion
impaired esthetics

## Introduction

Amelogenesis imperfecta (AI) is a hereditary disorder that includes a group of heterogeneous conditions where the structure and clinical appearance of the enamel is primarily affected.<sup>1-5</sup> Generally, both the primary and permanent teeth are involved, appearing smaller, discolored, and with irregular surfaces, and subject to accelerated wear. The dentin and roots are usually normal, even though pulp-chamber enlargement and pulp calcifications have been reported.<sup>1</sup> The most common clinical findings related to AI are listed in Table 1.<sup>6,7</sup>

Normal enamel formation can be divided into three stages: translation and secretion of the extracellular matrix, mineralization of the matrix, and maturation of the enamel. AI results from a disruption in at least one of these stages, with the diversity of phenotypes reflecting the stage at which the disruption occurred.<sup>5-8</sup> In 1956, Darling proposed the first AI classification, identifying five phenotypes.<sup>9</sup> Since then, several authors have tried to order the different expressions of the genetic mutation, which has led to 12 different classifications.<sup>1,3,10</sup>

The most widely used classification, based on the enamel appearance and the hypothesized developmental defects, was proposed by Witkop in 1988 (Table 2), and revised by Nusier in 2004.<sup>10-12</sup> Four types are distinguished: hypoplastic, hypomaturation, hypocalcified, and hypomaturation-hypoplastic.<sup>1-3,10-13</sup>

## Type I: Hypoplastic

In this type of AI, the enamel is well mineralized but its thickness is reduced. Clinically, grooves and pits are visible on the enamel surface. The teeth are tapered towards the incisal/occlusal face, and the contact points are open. Radiographs show a local or generalized decreased amount of enamel thickness, which appears as a thin, radiodense shell on the clinical crowns, with flat or absent cusp tips. Type I, which represents 60 to 73% of all AI cases, corresponds to a defect in the formation of the enamel matrix.



## Type II: Hypomaturation

The enamel presents a normal thickness with a hard consistency and a mottled opaque, white to yellow-brown or red-brown appearance. The compromised enamel tends to chip away from the underlying dentin rather than wear off. Radiographs show that the enamel thickness is within the normal amount, but its density is comparable to that of the dentin. A defect in the removal of extracellular matrix protein is thought to be the cause of this phenotype, resulting in decreased mineral deposition and increased matrix retention. Hypomaturation variants of AI represent 20 to 40% of all cases.

## Type III: Hypocalcified

This phenotype shows pigmented, softened, and easily fractured enamel, which also appears opaque or chalky. Radiographs show normal enamel thickness (immediately after tooth eruption), but with a density that is even less than the dentin. Hypocalcified variants of AI represent 7% of all cases.

## Type IV: Hypomaturation-hypoplastic

Affected patients present with the body and pulp chamber of their teeth enlarged, and with the floor of the pulp chamber and furcation positioned more apically, creating posterior teeth with a long trunk and shorter roots.

Each of the main types can be divided into subgroups based on the inheritance pattern (autosomal dominant, autosomal recessive, sex-linked, etc).<sup>14-16</sup>

**Table 2** Classification of amelogenesis imperfecta proposed by Witkop (1988)

Type I	Hypoplastic
Type IA	Hypoplastic, pitted autosomal dominant
Type IB	Hypoplastic, local autosomal dominant
Type IC	Hypoplastic, local autosomal recessive
Type ID	Hypoplastic, smooth autosomal dominant
Type IE	Hypoplastic, smooth X-linked dominant
Type IF	Hypoplastic, rough autosomal dominant
Type IG	Enamel agenesis, autosomal recessive
Type II	Hypomaturation
Type IIA	Hypomaturation, pigmented autosomal recessive
Type IIB	Hypomaturation, X-linked recessive
Type IIC	Hypomaturation, snow-capped teeth, X-linked
Type IID	Hypomaturation, snow-capped teeth, autosomal dominant
Type IIIA	Autosomal dominant
Type IIIB	Autosomal recessive
Type IV	Hypomaturation-hypoplastic with taurodontism
Type IVA	Hypomaturation-hypoplastic with taurodontism, autosomal dominant
Type IVB	Hypoplastic-hypomaturation with taurodontism, autosomal dominant

From a clinical point of view, practitioners would benefit from a classification which defines the types of AI in a way that distinguishes when the enamel is sound enough to be preserved with adhesive techniques.<sup>17-25</sup> Unfortunately, such a classification does not exist, although recently some researchers have begun studying this fundamental aspect.<sup>1,15</sup> Generally speaking, how-



**Fig 1** A 24-year-old Caucasian female patient whose facial clinical examination revealed an increased lower third facial height, a convex profile, and a high smile line.

ever, it could be assumed that when the enamel is hard, even though reduced in thickness (as in the hypoplastic type), it may respond to bonding.

In this article, the treatment of a 24-year-old Caucasian female patient affected by the hypoplastic type of AI is discussed, for whom a multidisciplinary approach was necessary, including orthodontics, periodontics, and restorative dentistry. Only adhesive techniques with minimal (if any) tooth preparation were implemented. At the completion of the treatment, the patient was very satisfied with the esthetic result, while the clinician was very happy with the biological success (maximum tooth-structure preservation was obtained). The 5-year follow-up confirmed that the treatment choice had been correct and raised the question of whether aggressive treatments should be considered as a best option to treat all patients affected by AI.

### Case presentation

A 24-year-old Caucasian female patient came for an initial consultation in 2007. She was aware of her discolored and irregular-shaped teeth and was determined to improve the appearance of her smile. She also complained of generalized chronic tooth sensitivity. Her medical history was noncontributory, and she was not a smoker.

The facial clinical examination revealed an increased lower third facial height and a convex profile (Fig 1).

A Class II Angle molar relationship, Division 1 malocclusion, was evidenced on both right and left sides, corresponding to a skeletal Class II malocclusion, and a mandibular deficiency. The patient also had a constricted maxillary arch, leading to a bilateral posterior crossbite. Intraorally, the clinical crown height was reduced and the thin biotype gingival presented inharmonious





**Figs 2 (a to d)** Intraoral views and casts demonstrated the maxillary constriction, the bilateral lateral crossbite and Class II Angle Division 1 malocclusion. An open bite was also present. The surfaces of the teeth were irregular, with the clinical crowns not completely developed, leading to multiple diastemata.

levels, very visible when smiling (severe gummy smile). At the level of each tooth, the enamel was thin and rough, but still hard. However, the dentin was exposed in several areas, explaining the patient's generalized tooth sensitivity (Fig 2).

Due to the irregular tooth shape, multiple diastemata were evident. An open bite was also present as a result of a 7 mm overjet and no vertical overlap (overbite). In addition, a dysfunctional swallowing was identified. Even though the patient's oral hygiene was considered poor, her motivation and efforts to



**Fig 3** The panoramic radiograph. While the roots, dentin and pulp anatomies were within the normal ranges, a generalized reduced thickness of the enamel, and several impacted teeth with crowns not fully developed, confirmed the diagnosis of hypoplastic type AI.



**Fig 4 (a and b)** Post-orthodontic intraoral view. Even though the tooth position was not ideal, the orthodontic treatment was concluded upon the patient's request. Fortunately, the palatal expansion already obtained was sufficient to proceed with the restorative phase.

achieve proper oral hygiene were high. No pocket depth deeper than 3 mm was recorded, regardless of the generalized gingivitis (presence of bleeding upon probing). At the time of first evaluation, defective restorations and caries lesions were present on several teeth. Emergency endodontic treatments had been performed on teeth 26 and 46 by her previous dentist. The radiographic examination showed a compromised enamel layer (thin to absent) on all the crowns of erupted teeth. In addition,



**Fig 5** After the orthodontic therapy, the overjet was reduced to 5 mm but the open bite was still present. Additive restorations involving facial and palatal veneers were planned to establish the final anterior contact points.

the third molar and the maxillary right second molar were impacted, with their crowns partially developed. However, the roots of all the teeth appeared normal in size and shape (Fig 3).

Considering these clinical findings, a diagnosis of hypoplastic type AI was made, based on Witkop's classification.<sup>11</sup>

Due to the patient's severe malocclusion, orthodontic therapy was recommended to expand the maxillary arch and achieve better posterior occlusal contacts prior to starting the reconstructive phase of the therapy. A removable appliance was used for 1 year to expand the palate. A Class I Angle molar relationship and better canine positioning were achieved on both right and left sides (Figs 4 and 5). However, the maxillary anterior teeth were not intruded during the orthodontic treatment, and the esthetic concern regarding the gummy smile remained. Several esthetic parameters, in particular the final incisal edge position, were evaluated by using phonetics first, followed by a mock-up.

When pronouncing the letter "e", the space between the upper and lower



**Fig 6 (a and b)** The patient's gummy smile was significant, and impossible to resolve without affecting the periodontal status of her teeth. A compromise was achieved whereby the gummy smile was reduced by a minor crown-lengthening procedure, but not completely corrected. The patient understood the limitations involved and agreed to a more conservative procedure.

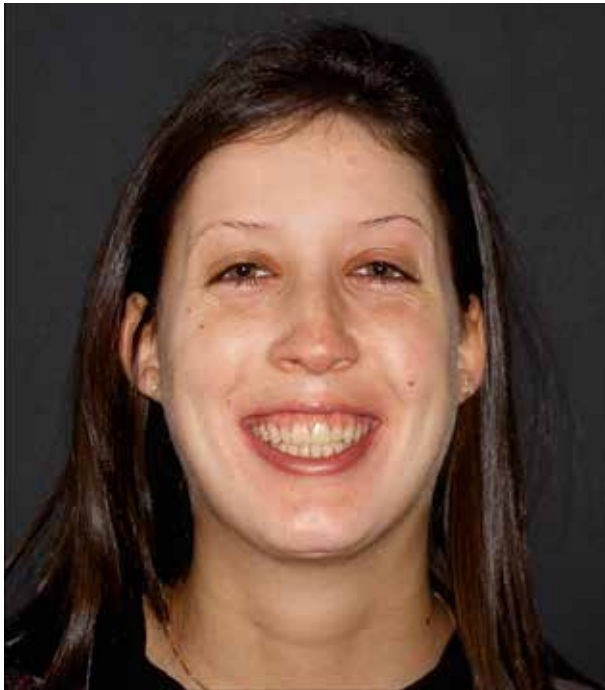
lips should ideally be occupied almost entirely by the maxillary incisors, especially in a young female. This patient's incisors only occupied about 40% of the interlabial vertical space (Fig 6). It was decided to retain the length of the right central incisor as a reference for the final incisors' edges, while lengthening the teeth in the cervical direction to partially reduce the gummy smile.

However, the gingival contours and dental proportions also appeared disharmonious in the posterior quadrants. To correct the short clinical crowns, the posterior teeth could have been lengthened in the occlusal direction by increasing the vertical dimension occlusion (VDO) and lowering the occlusal plane. However, based on the patient's facial height and the existing anterior open bite, it was considered inopportune to increase the VDO, and a more invasive solution was chosen (crown

lengthening, and more invasive occlusal preparation for the posterior teeth). To obtain the patient's consensus for the crown-lengthening procedure and to correctly evaluate its extent, full arch impressions were taken and study models were mounted in maximum intercuspal position (MIP) using a facebow transfer and a semi-adjustable articulator.

A full-mouth wax-up was requested and the laboratory technician was asked to extend the wax on the cervical margins, increasing the height of the original crowns. Two silicon keys were fabricated on the waxed-up models and filled with a tooth-colored, autopolymerizing composite material (CoolTemp, Coltène). Two full-mouth mock-ups were obtained directly in the mouth. Due to the mock-up extending above the gingival margins, the final position of the clinical crowns after surgery was visualized. With the mock-up in place, the





**Fig 7** Full-mouth direct mock-up. The tooth-colored autopolymerizing composite resin material overlapped the gingiva. The patient was very pleased with the esthetic outcome, and the clinician was able to evaluate the planned outcome after crown lengthening.



**Fig 8 (a and b)** Before and after crown lengthening (6-months follow-up), the height of the crowns was then more compatible with the restorative requirements.

patient was able to evaluate her future smile, and approved the clinician's esthetic project (Fig 7).

With the patient's approval, the crown-lengthening surgery was performed but kept to a minimum. It extended from teeth 6 to 6, maxillary and mandibular. The bone removal was limited to the buccal aspect and did not involve the interproximal areas,<sup>26</sup> especially at the level of the first molars, where the furcations were very close. A 6-month healing time followed before any restorative treatment took place, to allow sufficient time for the maturation and stability of the gingival tissues (Fig 8).<sup>27</sup>

New study models were then obtained and mounted, using a facebow transfer in a semi-adjustable articulator. The technician waxed up the two arches again, covering the occlusal irregular surfaces without increasing the VDO. Another maxillary key was fabricated. A new full-arch maxillary mock-up was delivered. After evaluating the esthetic outcome (incisal edges and occlusal plane position), the mock-up was used as a reduction key. Through the mock-up, depth cuts of 1.5 mm were made. In the same visit, all the maxillary premolars and the first molars were prepared for veneers/onlays (Fig 9).

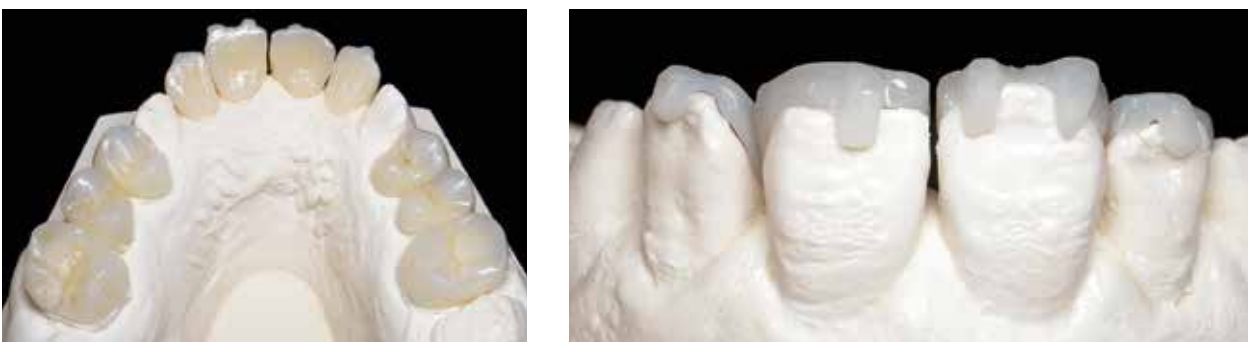


**Fig 9** The posterior veneer/onlays were prepared using the mock-up as a guide. **(a)** 1.5 to 2 mm depth of cut. The maxillary arch was treated at the same time. The preparation of palatal composite resin veneers on the incisors did not require any tooth preparation. **(b)** The immediate dentin sealing of any exposed dentin was done under a rubber dam.

During the same appointment, the maxillary anterior teeth were restored with palatal veneers. It was decided not to restore the palatal aspect of the two canines, since the final contact point with the antagonistic teeth would have been achieved by the mandibular restorations, eliminating the need to bulk the palatal surfaces of the canines (Fig 10).

Once the mock-up remnants were eliminated, the preparations were finalized using a rubber dam in order to allow

the sealing of the dentin before the impression under the most favorable moisture-control conditions. To immediately seal the dentin, each area of exposed dentin was roughened using a fine grit diamond bur (25 to 50  $\mu$ m). The enamel was etched for 30 to 45 s. The dentin was treated for only 15 s, then sealed immediately, using a 3-step etch-and-rinse adhesive (Optibond FL, Hawe-Kerr).<sup>28</sup> Finally, a thin layer (<1 mm) of flowable composite resin (Tetric Flow, Ivoclar Vi-



**Fig 10** Composite veneer/onlays and palatal veneers (Signum, Heraeus Kulzer) were fabricated on the master model at the same time. **(a)** Notice that the 2 canines remained unrestored. **(b)** Incisal hooks were used for the palatal veneers to help position the restorations.



**Fig 11** The palatal aspect of the maxillary anterior teeth presented an irregular enamel. Before the final impression for the palatal veneers, the surfaces were etched and sealed with bond and flowable composite resin to smooth their aspect.

vadent) was placed to smoothen the preparation surfaces.<sup>28-33</sup> The cervical margins remained located in enamel, 2 mm supragingivally. The interproximal contacts were slightly opened using thin diamond strips.

The preparation protocol was slightly different for the incisors because the teeth did not present exposed dentin on their palatal aspects, although their enamel surfaces were extremely irregular. The palatal surface irregularities were cleaned using airborne-particle abrasion (CoJet, 30  $\mu$ m aluminum oxide particles, 3M ESPE), acid-etched for 30 to 45 s with 35% phosphoric acid, followed by the application of bond (Optibond FL, Kerr). A thin layer (<1 mm) of flowable composite resin (Tetric Flow) was used to cover the preparation surfaces. Finally, the incisal edges were smoothened by removing the unsupported enamel prisms (Fig 11).

Before the final impression, the surfaces coated with composite resin were polymerized under glycerin gel to prevent any interactions between the oxygen-inhibited resin layer and the impression material.<sup>34</sup> A simple anterior bite registration was required, since the VDO was maintained by the second molars that had been left unrestored. The patient was invited to bite down on the registration material and her MIP was recorded.

To provisionalize the posterior teeth, a quick albeit unesthetic solution was preferred. After isolating the preparations with glycerin gel, a one-piece, soft provisional composite resin material (Fermit, Ivoclar Vivadent) was inserted bilaterally and the patient was again invited to bite in order to record her MIP. The resin was adjusted and then light-cured without cementation.<sup>35</sup> No provisional restorations were delivered on the anterior teeth.



**Fig 12 (a and b)** Details of the bonding of the maxillary restorations, and the final outcome. At this stage of the treatment, the occlusal plane was not in harmony with the incisal edge position (reverse smile). With the delivery of the facial veneers at the level of the maxillary anterior teeth, the harmony was then obtained.

After 1 week, the palatal veneers and the posterior overlays were bonded under rubber-dam isolation, following a similar protocol previously described by several authors (Fig 12).<sup>30,35-39</sup>

The hybridized dentin was abraded (CoJet), the surrounding enamel was etched (37% phosphoric acid) for 30 to 45 s, and 3 coats of organic silane (Monobond-S, Ivoclar Vivadent) were applied for 1 min. The preparation at the level of each tooth was completed with a layer of hydrophobic light-curing bonding resin (Optibond FL), which was applied but not cured. Simultaneously, the inner surfaces of the restorations were conditioned by airborne-particle abrasion (CoJet). Three coats of silane (Monobond-S) were then applied for 60 s and well dried with oil-free compressed air. Finally, a thin layer of bond (Optibond FL) was applied but not cured.<sup>36</sup> The restorations were bonded one by one using a preheated (Calset, AdDent) restorative light-curing composite resin (Miris 2, Coltène). The composite was directly applied onto the tooth before seating each restoration. A firm digital pressure on the occlusal sur-

face was used, and the final sitting of the restorations was obtained with the aid of a plastic-coated ultrasonic tip (Luting tip, EMS). Due to the presence of composite resin hooks at the level of the incisal edge of the palatal veneers, their insertion and proper positioning were easily handled.<sup>39</sup> For each restoration, a polymerization of 1 min per surface was adopted, with attention being paid not to heat the underlying pulp (Fig 12).

The rehabilitation of the maxillary anterior teeth was completed by means of facial veneers (sandwich approach, or bilaminar technique), as described by Vailati and Belser.<sup>38-40</sup> Two additional appointments were made to deliver 6 laminate ceramic veneers (from canine to canine). The ceramic veneer preparation followed standard protocols, developed and described in detail by various authors (Fig 13).<sup>30,33,37,38,42-44</sup> Due to the already tested additive mock-up, very little tissue was sacrificed. The reduction of the incisal edges by 1.5 mm was, in fact, obtained only by removing the length added by the palatal veneers.<sup>38</sup> A light chamfer was created fol-





**Fig 13** The bonding of the maxillary ceramic veneers was achieved under rubber dam isolation, starting with the central incisors and proceeding, 2 by 2, to the lateral incisors and canines on each side.

lowing the gingival margin profile, as the underlying teeth did not need to be hidden. The interproximal contact points, already opened during the preparation of the palatal veneers, were simply smoothed with a fine diamond strip, together with the sharp angles (Fig 13).

Immediate dentin sealing was then performed, as previously described. The final impression was carried out using polyvinyl siloxane materials (Virtual, Hawe-Kerr) and provisional veneers

were fabricated using the mock-up silicon key. The retention of the splinted provisional veneers relied on the polymerization shrinkage of the provisional composite material and the presence of minimal interproximal excesses.

The bonding of 6 feldspathic veneers (Willi Geller Creation, Jensen) was carried out 2 weeks later, following the protocol first described by Pascal Magne (Fig 14).<sup>33,42,43</sup>

After completing the restoration of the maxillary arch (except for the second molars), further appointments were scheduled to restore the mandibular posterior teeth. This time quadrant dentistry was preferred, rather than restoring the mandibular arch at the same time. Four appointments were necessary, with the task being facilitated by the presence of the contralateral posterior support that was maintaining the VDO, and the correct occlusal relationship (MIP).

After restoring the posterior quadrants, the 6 mandibular anterior teeth were restored. A mock-up guided the final preparation for 6 ceramic facial veneers (Fig 15).



**Fig 14 (a and b)** Minimally invasive preparations were necessary to deliver a 1.5 mm average thickness for the restorations. Preparations and cementation were done under rubber dam isolation and the cervical margins were established 2 mm supragingivally at the level of the cervical enamel.





During the immediate dentin sealing of the exposed dentin, to correct the irregular facial surfaces and allow for the fabrication of veneers of equal thickness, the vestibular surfaces of the teeth were bulked with a layer of composite (Miris 2).<sup>43</sup> Finally, the lingual aspect was restored with direct composite restorations. After 2 weeks, 6 feldspathic veneers were delivered, following the same adhesive protocol previously described (Fig 16).



**Fig 15** The treatment ended by restoring the mandibular anterior teeth using ceramic veneers on the buccal side and direct composite restorations on the lingual aspect. The preparations of the veneers were guided by the mock-up to obtain an equal thickness for feldspathic ceramic (about 1 to 1.5 mm). Direct composite resin restorations were implemented before the final impression.



**Figs 16 (a to c)** Bonding of 6 ceramic veneers and follow-up. Note the perfect gingival integration at the 15-day follow-up.



**Fig 17 (a to e)** Final result at 5-year follow-up. Although the gummy smile was still present, a better esthetic balance was achieved between the white and pink components of the patient's smile.



At the completion of treatment, the patient entered into a recall program of 3 to 4 months with a dental hygienist. The treatment ended in January 2010, and during the 5-year observation time only 3 teeth presented clinical complications. Specifically, teeth 27 and 47, which had been previously restored with direct composite resin restorations, presented recurrent car-

ies lesions and were restored again in the same manner (with direct composite resin restorations). In addition, the patient had been severely attacked in 2014 and suffered bruising and multiple wounds on her face and body. Intrabuccally (only) a necrosis was then diagnosed on tooth 43, and root canal treatment was performed on this tooth. Otherwise, the tooth sensitivity



was greatly reduced and, due to the smoother tooth surfaces, oral hygiene was facilitated, with an overall improvement in the patient's periodontal status (eg, no bleeding at probing). The composite restorations aged without macroscopic signs of excessive wear, except the loss of the glossy surfaces, as is routinely observed in cases of extensive composite restorations. The restoration margins did not present any discolorations or infiltrations (Fig 17).

Finally, the patient was thrilled with the esthetic result, which had changed her social life incredibly (Fig 18).



**Fig 18** The patient was thrilled with the final esthetic outcome that was achieved with minimal tooth removal (biological success).

## Discussion

A correct diagnosis and adequate treatment planning are fundamental to obtaining a satisfactory treatment, especially when a multidisciplinary approach is required.<sup>7,18,19,21,25</sup> For patients affected by AI, the treatment presents an even higher level of complexity, since clinicians are not only overwhelmed by the severity of the tooth destruction, but are also unsure about when and how to treat these patients.<sup>17</sup> In addition, since it is commonly agreed that the altered enamel layer should be completely removed, full coverage remains the treatment of choice.<sup>1,17,18,22,24</sup> As a full-mouth conventional rehabilitation is too aggressive, however, it is generally postponed until more spontaneous degradation of the tooth structure becomes evident. Even though this approach avoids early mutilation of the affected patient's dentition, it does not stop the progression of tooth structure loss because, unprotected by the alter-

ated enamel, the clinical crowns show an accelerated wear.

Adhesive techniques, already implemented in the treatment of young patients affected by other types of tooth destruction (eg, erosion), may be a valid option instead, ie, to intercept these patients earlier with a minimally invasive approach.<sup>38-40</sup>

When a full-mouth adhesive rehabilitation is being considered, clinicians should know the specific type of AI, as not all forms respond favorably to enamel bonding; much depends on the histological structure of the enamel and dentin of affected patients. AI can manifest in different ways and affect dental enamel differently. There is therefore no one definitive treatment protocol. The main difficulty lies in the diagnosis itself, which should be mainly based on macroscopic findings as there is growing acceptance that a classification of (inherited) enamel defects based primarily or



exclusively on phenotype (appearance) is problematic and can lead to misclassification of cases.<sup>14</sup> For this reason, the pattern of inheritance, the mutation involved, the expression of matrix proteins, and the biochemical changes associated with the mutations appear to be important discriminators.<sup>15</sup>

Genetic diagnosis, however, is presently only a research tool. In the future, genetic testing will be available to identify the specific mutation that causes an inherited disease in a given family.<sup>16</sup> Dental professionals will be able to order these tests; first, to affirm a positive diagnosis for each member of a family and, second, to enable more accurate treatment decisions to be made. For instance, as was previously mentioned, not all forms of AI respond favorably to enamel bonding, the success of which may depend on which specific genes are affected.<sup>2,12,15</sup> For example, researchers have found that mutations in the *KLK4* gene result in enamel malformations that do not respond to etching and bonding, while those caused by defects in the *enamelin* or *amelogenin* genes do.<sup>41</sup> Amelogenin is the main protein associated with the formation and characteristics (eg, shape and thickness) of dental enamel. If the prime defect is in the amount of enamel matrix produced, the enamel will be hard, normally translucent, and not subject to significant attrition. It might then respond to etching and bonding. In addition, acid etching and examination with scanning electron microscopy<sup>42</sup> have suggested that the presence of three different patterns in hypoplastic type AI was evidence of normal prism structure, as described by Silverstone

et al.<sup>43</sup> Further, microanalytical studies with quantitative electron microscopy of the calcium concentrations have shown that there were no significant differences in the effects of acid etching between hypoplastic type AI and healthy control teeth.<sup>44-46</sup> The underlying dentin structure variations are also considered to be comparable to normal teeth. Those cases with enamel that is reasonably hard (ie, less hypomineralized) and thin (ie, more hypoplastic) lend themselves fairly readily to the use of adhesive procedures.

By contrast, in some individuals and families presenting AI, the phenotype may be predominantly or exclusively hypomineralized.<sup>16</sup> In these cases, etch patterns of clinical variants of AI enamel may be altered and may not produce a good match to normal enamel.<sup>42-44</sup> Additionally, the morphological pattern of dentin in hypocalcified AI is relatively similar to sclerotic dentin, which responds to acid-etch conditioning differently to the way normal dentin responds.

Thus, given the complexity of enamel formation and the variability of the genetic expression of AI, it is probably realistic to routinely base dental treatment plans on macroscopic observations until such time as the molecular and protein pathways of enamel formation have been elucidated, and reliable genetic tests are available.

The patient described in this article is an example of clinically acceptable results, considering the lack of bonding failure after 5 years. These observations also accord with case reports previously published by other authors.<sup>16,17,19,46</sup> However, since the affected enamel remains a questionable substrate for



bonding, early failure of the restorations should be considered a risk of treatment in some patients.

Further, multiple reinterventions on the same tooth should be anticipated during the patient's life. However, due to the available adhesive techniques and pulp vitality, tooth structure loss could be kept to a minimum, especially for the anterior teeth, with their triangular shape, where the avoidance of full crowns will preserve a significant amount of tooth structure (eg, mesially and distally). The adhesive treatment proposed in this article – the sandwich approach (or bilaminar technique) – consisted in delivering two veneers, one facial and one palatal.<sup>38-40</sup> Thanks to two different paths of insertion, there was no need to remove the undercuts and additional healthy tooth structure.

We are of the opinion that clinicians should try using adhesive techniques more often for this heterogeneous population of patients, especially considering that in some AI variants, the altered enamel still represents an acceptable substrate for bonding.

## Conclusions

AI is a complex hereditary disorder that affects the enamel in different ways. One common finding, however, is the psychological struggle of the affected patients, which should influence clinicians to address the dental problem as soon as possible. On the other hand, decision-making regarding when and how to start treatment is a challenge, considering the literature only provides case reports with limited follow-ups.

Regarding the highly variable data, adhesion on affected enamel cannot be considered to be as reliable as other restorative treatments. Nevertheless, in our opinion, adhesion still remains the first option in order to achieve an early, minimally invasive intervention. AI patients should be made aware that they will need to have their restorations replaced several times in their lives. However, we are of the opinion that in the long term, fewer biological complications (such as loss of vitality) will be encountered if an early adhesive rehabilitation is carried out.

In this clinical report, restorative dentistry was performed on a 24-year-old Caucasian female patient suffering from hypoplastic type AI, using only adhesive procedures. Due to the complex needs of the patient, an interdisciplinary approach was followed, involving orthodontics, periodontics, and restorative dentistry. The aim of the treatment was to immediately protect the remaining tooth structure while improving the esthetics and enhancing the masticatory function of the patient. A full-mouth adhesive rehabilitation was achieved by means of direct composite restorations, veneer/onlays and facial/palatal veneers. No elective endodontic therapy was necessary for restorative purposes. The patient was thrilled with the result, and due to the minimal invasiveness of the overall treatment, all the teeth had retained their vitality even at the 5-year follow-up (except for a traumatic necrosis on tooth 43).

The esthetics, mechanics, and biological success were achieved and maintained. The bond to the enamel did not show signs of degradation (eg, discoloration or infiltration), which is encouraging to clinicians as it shows that





adhesive techniques may be reliable even in the presence of a compromised enamel layer.

## Acknowledgements

Gratitude is expressed to Dr Sandra Mongaillard, who performed the orthodontics, and to Dr Anne-Marie Neiderud, who performed the crown lengthening. Special thanks to Samuel Schwab (Lys Dental, Lausanne) for his outstanding laboratory work.

## Conflict of interest declaration

The authors certify that they have no commercial associations that might represent a conflict of interests in connection with this article.

## References

1. Crawford PJ, Aldred M, Bloch-Zupan A. Amelogenesis imperfecta. *Orphanet J Rare Dis* 2007;2:17.
2. Poulsen S, Gjørup H, Haubek D, et al. Amelogenesis imperfecta – a systematic literature review of associated dental and oro-facial abnormalities and their impact on patients. *Acta Odontol Scand* 2008 Aug;66:193–199.
3. Bsoul SA, Flint DJ, Terezhalmay GT, Moore WS. Amelogenesis imperfecta. *Quintessence Int* 2004;35:338–339.
4. Canger EM, Celenk P, Yenisey M, Odyakmaz SZ. Amelogenesis Imperfecta, hypoplastic type associated with some dental abnormalities: a case report. *Braz Dent J* 2010;21:170–174.
5. Ayers KM, Drummond BK, Harding WJ, Salis SG, Liston PN. Amelogenesis imperfecta – multidisciplinary management from eruption to adulthood. Review and case report. *N Z Dent J* 2004;100:101–104.
6. Seow W K. Clinical diagnosis and management strategies of amelogenesis imperfecta variants. *Pediatr Dent* 1993;15:384–393.
7. Malik K, Gadhia K, Arkutu N, McDonald S, Blair F. The interdisciplinary management of patients with amelogenesis imperfecta – restorative dentistry. *Br Dent J* 2012;212:537–542.
8. Oliveira IK, Fonseca Jde F, do Amaral FL, Pecorari VG, Basting RT, França FM. Diagnosis and esthetic functional rehabilitation of a patient with amelogenesis imperfecta. *Quintessence Int* 2011;42:463–469.
9. Darling AI. Some observations on amelogenesis imperfecta and calcification of the dental enamel. *Proc R Soc Med* 1956;49:759–765.
10. Witkop CJ, Sauk JJ. Hereditary enamel defects. In: Stewart RE, Prescott GH (eds). *Oral facial genetics*. St Louis: CV Mosby, 1976:151.
11. Witkop CJ Jr. Amelogenesis imperfecta, dentinogenesis imperfecta and dentin dysplasia revisited: problems in classification. *J Oral Pathol* 1988;17:547–553.
12. Nusier M, Yassin O, Hart TC, Samimi A, Wright JT. Phenotypic diversity and revision of the nomenclature for autosomal recessive amelogenesis imperfecta. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2004;97:220–230.
13. Gadhia K, McDonald S, Arkutu N, Malik K. Amelogenesis imperfecta: an introduction. *Br Dent J* 2012;212:377–379.
14. Aldred MJ, Savarirayan R, Crawford P J. Amelogenesis imperfecta: a classification and catalogue for the 21st century. *Oral Dis* 2003;9:19–23.
15. Wright JT. The molecular etiologies and associated phenotypes of amelogenesis imperfecta. *Am J Med Genet A* 2006;140:2547–2555.
16. Wright JT, Torain M, Long K, et al. Amelogenesis imperfecta: genotype-phenotype studies in 71 families. *Cells Tissues Organs* 2011;194:279–283.
17. Elderton R, Nuttall N. Variation amongst dentists in planning treatment. *Br Dent J* 1983;154:201–206.
18. Toksavul S, Ulusoy M, Türkün M, Kümloglu O. Amelogenesis imperfecta: the multidisciplinary approach. A case report. *Quintessence Int* 2004;35:11–14.
19. McDonald S, Arkutu N, Malik K, Gadhia K, McKaig S. Managing the paediatric patient with amelogenesis imperfecta. *Br Dent J* 2012;212:425–428.
20. Sabatini C, Guzmán-Armstrong S. A conservative treatment for amelogenesis imperfecta with direct resin composite restorations: A case report. *J Esthet Restor Dent* 2009;21:161–169; discussion 170.
21. Varela Morales M, Botella Perez JM, Jiménez Garcia J, Varela PG. Interdisciplinary treatment of a patient with amelogenesis imperfecta. *J Clin Orthod* 2010;9:553–559; 562.
22. Ağaçkiran E, Tümen EC, Celenk S, Bolgöl B, Atakul F. Restoring aesthetics and function in a young boy with hypomature amelo-



- genesis imperfecta: a case report [epub ahead of print 21 Sept 2010]. ISRN Dent doi:10.5402/2011/586854.
23. Doruk C, Ozturk F, Sari F, Turgut M. Restoring Function and Aesthetic in a Class II Division 1 Patient with Amelogenesis Imperfecta: A Clinical Report. *Eur J Dent* 2011;5:220–228.
  24. Chamarthi V, Varma BR, Jayanthi M. Amelogenesis imperfecta: a clinician's challenge. *J Indian Soc Pedod Prev Dent* 2012;30:70–73.
  25. Khodaeian N, Sabouhi M, Ataei E. An interdisciplinary approach for rehabilitating a patient with amelogenesis imperfecta: a case report [epub ahead of print 16 Aug 2012]. *Case Rep Dent* doi:10.1155/2012/432108.
  26. Caudill R, Chiche GJ. Establishing an esthetic gingival appearance. In: Chiche GJ, Pinault A (eds). *Esthetics of fixed prosthodontics*. Chicago: Quintessence Publishing, 1994:177–198.
  27. Olsen CT, Ammons WF, van Belle G. A longitudinal study comparing apically repositioned flaps, with and without osseous surgery. *Int J Periodontics Restorative Dent* 1985;5:10–33.
  28. Magne P, So WS, Cascione D. Immediate dentin sealing supports delayed restoration placement. *J Prosthet Dent* 2007;98:166–174.
  29. Bertschinger C, Paul SJ, Lüthy H, Schärer P. Dual application of dentin bonding agents: effect on bond strength. *Am J Dent* 1996;9:115–119.
  30. Paul SJ, Schärer P. The dual bonding technique: a modified method to improve adhesive luting procedures. *Int J Periodontics Restorative Dent* 1997;17:536–545.
  31. Magne P, Douglas WH. Porcelain veneers: dentin bonding optimization and biomimetic recovery of the crown. *Int J Prosthodont* 1999;12:111–121.
  32. Magne P. Immediate dentin sealing: a fundamental procedure for indirect bonded restorations. *J Esthet Restor Dent* 2005;17:144–154; discussion 155.
  33. Belser UC, Magne P, Magne M. Ceramic laminate veneers: continuous evolution of indications. *J Esthet Dent* 1997;9:197–207.
  34. Magne P, Nielsen B. Interactions between impression materials and immediate dentin sealing. *J Prosthet Dent* 2009;102:298–305.
  35. Rocca GT, Krejci I. Bonded indirect restorations for posterior teeth: from cavity preparation to provisionalization. *Quintessence Int* 2007;38:371–379.
  36. Rocca GT, Krejci I. Bonded indirect restorations for posterior teeth: the luting appointment. *Quintessence Int* 2007;38:543–553.
  37. Dietschi D, Spreafico R. Adhesive metal-free restorations. Quintessence Publishing, 1997.
  38. Vailati F, Belser UC. Full-mouth adhesive rehabilitation of a severely eroded dentition: the three-step technique. Part 3. *Eur J Esthet Dent* 2008;3:236–257.
  39. Vailati F, Belser UC. Palatal and facial veneers to treat severe dental erosion: a case report following the three-step technique and the sandwich approach. *Eur J Esthet Dent* 2011;6:268–278.
  40. Vailati F, Belser UC. Full-mouth adhesive rehabilitation of a severely eroded dentition: the three-step technique. Part 1. *Eur J Esthet Dent* 2008;3:30–44.
  41. Vailati F, Belser UC. Full-mouth adhesive rehabilitation of a severely eroded dentition: the three-step technique. Part 2. *Eur J Esthet Dent* 2008;3:128–146.
  42. Magne P, Douglas WH. Additive contour of porcelain veneers: a key element in enamel preservation, adhesion, and esthetics for aging dentition. *J Adhes Dent* 1999;1:81–92.
  43. Magne P, Belser UC. Novel porcelain laminate preparation approach driven by a diagnostic mock-up. *J Esthet Restor Dent* 2004;16:7–16; discussion 17–18.
  44. Gürel G. The science and art of porcelain laminate veneers. Chicago: Quintessence Publishing, 2003.
  45. Lindemeyer RG, Gibson CW, Wright TJ. Amelogenesis imperfecta due to a mutation of the enamel gene: clinical case with genotype-phenotype correlations. *Pediatr Dent* 2010;32:56–60.
  46. Hiraishi N, Yiu CKY, King NM. Effect of acid etching time on bond strength of an etch-and-rinse adhesive to primary tooth dentine affected by amelogenesis imperfecta. *International J of Paediatric Dent* 2008;18:224–230.
  47. Silverstone LM, Saxton CA, Dogon IL, Fejerskov O. Variation in the pattern of acid etching on human dental enamel examined by scanning electron microscopy. *Caries Res* 1975;9:373–387.
  48. Seow WK, Amaratunge A. The effects of acid-etching on enamel from different clinical variants of amelogenesis imperfecta: an SEM study. *Pediatr Dent* 1998;20:37–42.
  49. Sánchez-Quevedo C, Ceballos G, Rodríguez IA, García JM, Alaminos M. Acid-etching effects in hypomineralized amelogenesis imperfecta. A microscopic and microanalytical study. *Med Oral Patol Oral Cir Bucal* 2006;11:E40–43.
  50. Smith RN, Elcock C, Abdelatif A, Bäckman B, Russell JM, Brook AH. Enamel defects in extracted and exfoliated teeth from patients with Amelogenesis Imperfecta, measured using the extended enamel defects index and image analysis. [epub ahead of print 2 Sept 2008]. *Arch Oral Biol* 2009;54:S86–S92.