Interdisciplinary Care for a Patient with Amelogenesis Imperfecta: A Clinical Report

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Abstract

This manuscript describes an interdisciplinary approach over a period of 8 years combining surgical and prosthodontic treatment of a young patient diagnosed with hypocalcified-type amelogenesis imperfecta and anterior open bite. The treatment procedures included transitional restorations, orthodontic treatment, and maxillofacial surgery with a one-piece Le Fort I osteotomy, bilateral mandibular osteotomy, and genioplasty. The definitive prosthetic rehabilitation consisted of 28 zirconia-based ceramic single crowns restoring both esthetics and function. Photographs and radiographs associated with clinical evaluation were used in the maintenance period. Two-year follow-up revealed satisfactory results and no deterioration in the restorations.

Clinical report

A 10-year-old girl and her parents presented to the Pediatric Dentistry Department of the Hospices Civils of Lyon, France, with the chief complaints of dental sensitivity to hot and cold food and discolored teeth. The patient reported difficulty with mastication and poor self-image due to teeth appearance. No remarkable findings were identified in her medical record. There was no evidence of systemic disease, nutritional deficiency, or drug treatments that may have affected dentition structure during development. According to her parents, none of her first-degree family members (mother, father, sister) exhibited the same dental problems. The clinical examination showed a skeletal class II malocclusion with a severe mandibular

Amelogenesis Imperfecta (AI) is the expression of a heterogeneous inherited enamel disorder associated with mutations in many genes encoding enamel proteins such as amelogenin, enamelin, enamelysin, KLK4, WDR72, and FAM83H.1,2 This genetic expression affects the quantity and quality of tooth enamel in both primary and permanent dentition, with variable prevalence from 1/700 to 1/14,000.3 AI has been reported as an isolated or syndromic finding with an autosomal dominant, autosomal recessive, or X-linked inheritance.4 The AI enamel defects are highly variable and may be classified as hypoplastic, hypomineralized (hypomaturation and hypocalcification), or both.5 Major oral complications in patients with AI are rapid wear and compromised dental esthetics that may disrupt their social lives. Other associated clinical features include gingival hyperplasia, delayed dental eruption, tooth agenesis, pulp stones, progressive root and crown resorption, and short roots.5,7 Craniofacial features may also be present, acting as constricted maxillary arch, reversed mandibular curve of Spee, and anterior and posterior open bite occlusions. In addition, these patients may face nutrition problems due to dental sensitivity and loss of occlusal vertical dimension caused by attrition.5,9 The management of the most complex cases with severe malocclusion is a challenge for clinicians and usually requires an interdisciplinary approach.10,11 The use of all-ceramic systems in such cases is considered, but some concerns regarding their durability have been reported.12,13 The purpose of this clinical report is to describe the interdisciplinary management and fixed rehabilitation with computer-assisted design (CAD)/computer-aided manufacturing (CAM) zirconia-based ceramic crowns of a young girl suffering from AI, and the 2-year follow-up results.
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Orofacial Rehabilitation of a Patient with Al

Figure 1 Initial presentation at age 10. Note the severe mandibular retrognathism and the convex profile.

retrognathism, a 5-mm anterior open bite, and incompetent lips at rest (Fig 1). Oral hygiene was poor, with evidence of gingivitis due to severe plaque accumulation, particularly around the existing composite resin restorations on the mandibular incisors. The patient had a mixed dentition. All erupted teeth had a yellowish discoloration, with rough surfaces and irregular defects (Fig 2). Enamel was softer than normal and showed some signs of detachment from the dentin. Intraoral periapical radiographs revealed no missing teeth, a reduced enamel thickness, a lack of distinction in density between enamel and dentin, and a carious lesion on the mandibular right first premolar (Fig 3). Pulp chambers had normal shape, and no calcification was noted. The clinical and radiographic appearance suggested a hypocalcified form of Al. According to the Prosthodontic Diagnostic Index, the patient was classified as Class IV.

The treatment objectives were to reduce tooth sensitivity, attain ideal functional occlusion, and improve esthetic appearance. Because of the severity of the problem and the young age of the patient, an interdisciplinary consultation with a pediatric dentist, an orthodontist, and a prosthodontist was necessary. Specialists jointly evaluated the case and discussed treatment planning and the ultimate restorative requirements. Lateral cephalometric analysis showed a mandibular retrognathia (ANB 14°) and an accentuated inclination of the mandibular plane (FMA 35°). The diagnostic procedure on casts mounted in centric relation indicated that mandibular and maxillary surgery, after completion of facial growth, combined with orthodontic treatment, was needed to correct the skeletal imbalance. The proposed treatment was surgical-orthodontic management followed by complete mouth rehabilitation using all-ceramic crowns to stabilize the results and improve tooth form and color. Despite the length, cost, and difficulty, this option would provide an esthetic change and correct the patient’s convex facial profile and the malocclusion. She and her parents were informed about the treatment plan with its objectives and possible complications. The importance of oral hygiene, caries control, and cooperation was emphasized. The patient and her parents gave their informed consent. The initial therapy consisted of dietary counselling and oral hygiene instructions emphasizing the use of tempered water to rinse the mouth. During the first sessions, preformed stainless steel crowns (Ion; 3M ESPE, St. Paul, MN) were placed subgingivally on the first permanent molars following minimal slice preparation of the teeth. The main goal of these crowns was to protect
the dentin/pulp complex as well as to decrease the risk of loss of vertical dimension. Maxillary and mandibular incisors received preformed polycarbonate resin crowns (Ion). During this initial phase the child was monitored every 3 months to follow up the eruption of all permanent teeth. Her cooperation and hygiene were good, confirmed by the improvement of her
giving health. After eruption, the premolars, canines, and second molars were fitted with preformed crowns cemented with glass ionomer cement (GIC; Fuji I; GC Corporation, Tokyo, Japan; Fig 4). Pulp vitality was maintained for all the teeth except the mandibular right first premolar, which required endodontic treatment after caries excavation.

When the patient was 14 years, 8 months old, fixed orthodontic appliances were placed on preformed crowns to align and level the dentition of both arches, to achieve better arch coordination prior to surgery. The presurgical operative orthodontic treatment was achieved within 21 months. After bone growth completion, preoperative planning of orthognathic surgery was realized through the use of 3-D computer-assisted simulation. Visual treatment objectives of the orthodontic treatment could achieve an Angle Class I profile by a posterior impaction of the maxilla and an 8 mm advance at the mandible. Orthognathic surgical splints were produced using the computerized digital model simulation. The patient underwent a midline segmental split Lefort I osteotomy in the upper jaw with impaction in the posterior region. Additionally, a bilateral sagittal split osteotomy with genioplasty procedure was realized to advance
the mandible and correct the mandibular retrognathism. After the consolidation period, postsurgical orthodontic treatment was processed on old preformed crowns for 11 months. Meanwhile, a preformed polycarbonate resin crown on the maxillary left lateral incisor crown was damaged and replaced by a new one. The retrusive chin and convex profile improved (Fig 5). A Class I occlusion with both normal horizontal and vertical overlap was achieved (Fig 6). During the retention phase of treatment, a maxillary removable wraparound retainer was used, and a fixed wire retainer was bonded from canine to canine in the mandibular arch. The overall orthodontic treatment lasted 32 months.

Following orthodontic treatment completion, the prosthodontic treatment was initiated at the age of 18. First, diagnostic casts of the patient’s jaws with the preformed crowns were obtained and mounted on a semi-adjustable articulator SAM 2 (S.A.M. Praezisontechnik; GmbH, Munich, Germany) using centric relation record. All posterior teeth had the necessary minimum clinical crown length. Therefore, no crown lengthening seemed necessary; however, the patient had an asymmetrical gingival level between the central and lateral incisors and canines of the maxillary dental arch. Periodontal surgery to correct the gingival level with a 3-month healing period was proposed, but the patient rejected this option. A wax-up was generated for the realization of interim prostheses with the
Purpose of serving as a blueprint for the definitive restorations. At this point, the patient requested that the definitive crowns on the anterior maxillary teeth be slightly less triangular than the preformed crowns. Furthermore, the patient’s expectation was to achieve some resemblance to her sister who presented with a slight extruded and rotated maxillary left lateral incisor. Thus, the wax-up was modified according to the patient’s demand (Fig 7). Central incisors were equal in width but not in height due to the asymmetric gingival heights. The wax-up was duplicated, and a vacuum-formed matrix was made. Preformed crowns were removed, and teeth were prepared under local anesthesia for all-ceramic single crowns. All teeth were prepared with an occlusal reduction of 1.5 to 2 mm, an axial reduction of 1 mm, and a finish line located below the cementoenamel junction to lower the risk of secondary caries due to altered enamel. Tooth preparations were finished by rounding sharp angles (Fig 8). Interim prostheses with a mutually protected occlusal scheme were made chairside using the matrix and autopolymerized acrylic resin (Unifast Trad; GC America, Alsip, IL). They were temporarily cemented using a zinc oxide-eugenol cement (TempBond; Kerr Italia, Scafati, Salerno, Italy). The patient wore her interim prostheses for 3 months, allowing sufficient time for adjustments to validate esthetics and function. Adjustments of the incisal edge position of the two maxillary central incisors were made both according to the patient’s esthetic expectations, and to the fleshy and slightly asymmetrical lower lip (slightly higher on left side). The restorations were further adjusted to avoid any occlusal interference in protrusive and lateral excursions. Poly(vinyl siloxane) material (Express; 3M ESPE) was used for the master impressions in custom impression trays. The maxillomandibular relationship was registered using wax wedges (Moyco Industries Inc, Philadelphia, PA). The maxillary and mandibular working casts (New Fujirock; GC) were mounted on a semi-adjustable articulator SAM 2. The dies were scanned using the CAD system (Cercon; DeguDent, Hanau, Germany) to acquire the data for a 3-D virtual model. All zirconia frameworks were designed using CAD software with anatomical shape to guarantee sufficient support for the veneering porcelain, and a thickness of 0.5 mm (Fig 9). Pre-sintered zirconium oxide frameworks (Cercon HT; DeguDent) for each tooth were milled individually by CAM. The zirconia copings were sintered and then clinically fitted on the abutments with a film of silicone material (Fit Checker; GC). All copings showed a satisfactory marginal adaptation with a dental explorer (Fig 10). For the veneering process, the copings were covered with a manual layering technique using a ceramic material (Cercon Ceram-Kiss; DeguDent). The occlusion was constructed as mutually protected occlusion without eccentric contacts. Canines disengaged the posterior teeth during lateral movements. Protrusive guidance was evenly distributed across the maxillary and mandibular incisors. A trial evaluation of the ceramic before glazing enabled minor occlusal adjustments using a turbine and diamond burs with 30 to 40 µm grain size. No crowns were provisionally cemented before definitive cementation. The luting of the 28 single-tooth crowns was achieved with a resin-modified GIC (Fuji Cem 2; GC).

Both the patient and her parents were very satisfied with the esthetic and functional results despite the occurrence of interdental papillae recessions (Figs 11 and 12). In the follow-up, maintenance of oral hygiene was emphasized, and an occlusal night guard was prescribed to prevent the restorations from chipping; however, the patient declined to wear the occlusal guard at night. The patient was recalled for follow-up at 3, 6, 12, and 24 months. During follow-up appointments, the crowns were clinically evaluated for marginal adaptation, fracture, surface chipping, retention, color stability, and wear. The clinical evaluation was performed using tactile perception with a sharp explorer and visual inspection. Biological properties such as tooth sensitivity, secondary caries, and periodontal response were also evaluated. Radiographic and photographic evaluations were made at 1- and 2-year recall visits. Intraoral digital photographic evaluation included frontal view of the anterior teeth and occlusal views of the full maxillary and mandibular arches. Radiographic assessment included bitewing and periapical radiographs. Evaluation 2 years after treatment showed good stability of both orthodontic and prosthodontic results. This esthetic result has definitely affected the quality of life of the patient. Teeth showed no sensitivity, and the restorations exhibited no signs of complication. Marginal adaptations were good. No fracture of framework, chipping of veneered porcelain, or loss of retention was noted. There was no radiographic evidence of proximal caries (Fig 13). As for gingival response, an improvement of interdental papillae was observed (Fig 14), and no functional complaint was noted. The clinical evaluation according to a modified California Dental Association (CDA) ranking, as confirmed with high patient satisfaction, yielded an “excellent” ranking for the 28 zirconia-based ceramic single crowns.

Discussion

This clinical report demonstrates that timely diagnosis and individual approach is important for adequate treatment of patients with severe AI and anterior open bite. The interdisciplinary approach is essential for successful management of structural, esthetic, and functional issues. Numerous treatment modalities have been described for rehabilitation of AI patients: adhesive restorative techniques, overdentures, fixed dental prostheses, all-ceramic crowns, metal ceramic restorations, and inlay/onlay restorations. Treatment modality includes consideration of a patient’s age, severity of AI, orthodontic and maxillofacial needs, periodontal condition, financial implications for the patient’s family, and long-term prognosis. In this young patient, preformed crowns were placed as transitional treatment prior to orthognathic surgery, which should be performed after the completion of growth to prevent recurrence and interference with craniofacial development. The management by direct interim bonding resin composite was not used due to poorly mineralized and friable enamel. Additionally, interim prostheses should have been maintained in the mouth for several years (for 5 to 7 years) during the eruption of all permanent teeth and then throughout the orthodontic stage prior to surgery. Furthermore, prefabricated stainless steel crowns are reported to be the most effective and efficient restorations in managing tooth sensitivity and restoring primary and permanent molars in children.
The option of initially using interim prostheses based on wax-up was not considered in this case due to the patient’s young age, the presence of mixed dentition, and insufficient space to correct tooth shape before the orthodontic treatment. In addition, tooth preparations had to be minimized due to large pulps. Here, orthodontic and maxillofacial procedures were realized prior to the wax-up, since a satisfactory occlusal relation could be established with the preformed crowns. Alternatively, in more complex occlusal conditions, the wax-up may be performed prior to orthognathic surgery. This enables the placement of interim prostheses in the desired occlusal final relationship, prior to surgery, thus guiding the surgeon for proper intraoperative jaw positioning. In the present case, an alternative orthodontic treatment could have been proposed to the patient to avoid maxillofacial surgery. It consisted of intruding the posterior teeth in both arches and extruding the maxillary anterior teeth to close the open bite; however, the potential change was not considered sufficient with this option. In case of denial of orthodontic and surgical treatments, periodontal surgical procedures with bone reduction on molars would have been needed for improving the occlusal plane and the anterior open bite. This option would not allow maintaining posterior tooth vitality and correction of skeletal discrepancy to achieve a normal profile. For the above reasons, orthodontic and surgical corrections were accomplished for a better esthetic and functional result.

Complete crown coverage is commonly recommended for the definitive restorative procedure in most severe hypocalcified AI cases. Even though significant reduction of tooth structure is required, such restorations protect the dental tissues from further destruction due to brittle enamel structure. Adequate tooth preparation and appropriate choice of restorative materials are essential to limit biological complications like loss of pulp vitality and to ensure long-term success. All-ceramic zirconia-based restorations were selected because all-ceramic restorations show good success rates with better esthetics compared to metal ceramic restorations. Besides, all-ceramic materials hold other advantages including low plaque retention and optimum biocompatibility inducing favorable biological responses in the soft tissues. In particular, zirconia demonstrated the possibility of inducing an epithelial attachment to its surface. Recent improvements in CAD/CAM technology enable shaping the zirconium dioxide framework with anatomic form. Thus a layer of uniform veneering porcelain technology enable shaping the zirconium dioxide framework with anatomic form. Thus a layer of uniform veneering porcelain can be created throughout the framework, substantially reducing the risk of chipping, especially in the posterior area. In addition, chipping may be reduced by using a coefficient of thermal expansion of veneering porcelain similar to zirconia. For this patient, these recommendations were followed, and the complete mouth rehabilitation provided a mutually protected occlusion to prevent structural alteration of the ceramic in the posterior area during excursions. At the 24-month clinical short-term examination, no fracture or chipping problems were recorded in the 28 single crowns. This observation was in agreement with data from recent published studies on the clinical short/medium-term performance of zirconia-based ceramic single crowns. Besides the ceramic system selected, the choice of the luting agent is important because the ceramic fracture strength decreases as the restoration ages and the bond strength degrades in the oral cavity. Because of its high flexural strength, zirconia can be conventionally cemented just like metal ceramic crowns, without the need for any pre-treatment, or bonded with adhesive resin cements. For this young patient with vital teeth and retentive preparations, resin-modified GIC was chosen due to its lower technique sensitivity, its good long-term results, and its potential for fluoride release. Although this luting agent provides inferior bond strength compared with resin cements, in vitro studies have demonstrated an improved retention strength for zirconia crowns compared to zinc phosphate and GIC cements.

Conclusion

Management of AI patients is a long and complex process requiring a carefully established protocol generally extending over several years. Coordinated interdisciplinary procedures are critical for a successful outcome and patient satisfaction. Prosthodontic rehabilitation with zirconia-based restorations represents a promising alternative, especially in the posterior area, to preserve and stabilize weakened tooth substance; however, longer observation periods and randomized controlled trials with a large number of treatments are necessary to assess the long-term success of such restorations.

References