International Journal of Dental Sciences
www.dentaljournal.in
E-ISSN: 2663-4708, P-ISSN: 2663-4694
Received: 31-05-2025, Accepted: 01-07-2025, Published: 16-07-2025
Volume 7, Issue 3, 2025, Page No. 21-26

Restorative treatment of teeth affected with amelogenesis imperfecta: A systematic review

Dr. Agarwal Rashmi G[1], Dr. Bengude Vikas G[2], Dr. Akkinapally Meghana[3], Dr. Harikesavan Iyer[4], Dr. P Aravind[3]
[1] Department of Pediatric and Preventive Dentistry, Dr. D Y Patil Vidyapeeth's Dr. D Y Patil Dental College and Hospital, Pimpri, Pune, Maharashtra, India
[2] Former Head and Professor, Department of Pediatric and Preventive Dentistry, Dr. D Y Patil Vidyapeeth's Dr. D Y Patil Dental College and Hospital, Pimpri, Pune, Maharashtra, India
[3] Assistant Professor, Department of Conservative Dentistry and Endodontics, Tirumala Institute of Dental Sciences, Ditchpally, Nizambad, Telangana, India
[4] Department of Pedodontics and Preventive Dentistry, Cupident Pediatric and Multispeciality Dental Studio, Mumbai, Maharashtra, India

Abstract
Amelogenesis imperfecta is a clinical condition, which comprises developmental disorders that demonstrate alterations in the enamel. It represents a group of inherited disorders, which are clinically heterogeneous and exhibit tooth enamel defects in the absence of systemic manifestations. It affects both deciduous and permanent dentition giving a characteristic feature like thin and soft enamel, pitting over the enamel surface, discoloration of teeth and loss of normal function. Literature reports various restorative modalities used as a treatment to amelogenesis imperfecta to improve the overall quality of life of patients but the treatment remains a challenge for the dentists.

Keywords: Congenital enamel hypoplasia, hypoplastic type amelogenesis imperfecta, autosomal recessive amelogenesis imperfects, ADHCAI

Introduction

AI is an uncommon genetic disease affecting enamel. Deciduous teeth and permanent teeth are affected. It is very essential to identify the amelogenesis imperfecta cases from that of other enamel defects which are caused by environmental elements like fluoride, tetracycline [1] or traumatic injuries as these factors will affect a particular dentition rather than both the dentitions. For instance, literature reports experimental studies which have reported that molar incisor hypoplasia (MIH), affects selectively the first permanent molars and permanent incisors and are caused by either prenatal or early childhood exposure to endocrine disruptors [2]. AI displays a huge variety in its clinical presentation. Mutation in the genes is responsible for these changes. Few of these encode for proteins like amelogenin, enamelin or ameloblastin, few for the enzymes, cellular proteins while few for calcium carriers. Until today, no connection between genotype and phenotype has been set up through the literature [2]. The changes identified in the enamel are with respect to its width, microstructure or mineralization degree. Thus, these changes are reflected into the appearance of enamel.

According to Witkop's classification, there are 4 different types of AI: hypoplastic, hypomature, hypomineralized and hypomature with taurodontism structures [3]. Its clinical appearance can be strikingly unique between these different types. Hypoplastic AI also called as type I, represents quantitative alterations in the enamel. The overall thickness of the enamel is reduced which can be localised or generalized. The colour ranges from yellow to light brown and the surface of the enamel is rough with pits of varying sizes [4]. The hypomature AI also known as type II represents defect in the matrix degradation. Ideally the proteins in the enamel are degraded to achieve the final crystal but, in this type, the final crystal formation is hampered. The enamel appears white to brown in color without translucency. The thickness of the enamel is normal [4]. The hypomineralized AI also known as type III is the most severe form of all. Enamel colour ranges from dark yellow to brown and the teeth are extremely sensitive to changes in temperature. Radiographically, the density of enamel and dentin may appear same [4].

Patients experience decreased quality of life, have difficulty in social integration and suffer from loss of confidence [5]. Oral hygiene maintenance and thorough follow-ups are advised for AI patients. Hypo-mineralized type III shows a progressive alteration with time due to softness of enamel. This degradation can be restricted by providing bonded restorations & overall dental rehabilitation is required to improve the oral health in children. Since the surface of enamel is rough, it tends to accumulate more amount of dental plaque which eventually forms calculus leading to gingivitis and poor periodontal health. Hence, maintaining the gingival health also becomes a priority [6].

As patients with AI complains of pain while eating due to change in temperature, a proper tooth brushing cannot be accomplished in these patients due to sensitivity of the teeth. Conversely, hypoplastic form of AI principally present teeth while in hypomineralized type, local anesthesia is required even for dental scaling. When it comes to initiation of treatment; in mixed dentition, restoration must be done as soon as the teeth erupt in the oral cavity. Primary objectives of treatment should be conservation of tooth integrity and vitality [7]. Paediatric crowns can be effectively placed on first molars with minimal tooth preparation. This procedure is especially indicated when the teeth are painful and hypoplastic. With the advancement in techniques and increase in availability of various dental materials, many studies have shown that the use of GIC, composite resin veneers, porcelain veneers, SSC, lab-fabricated crowns, and over dentures can restore the affected teeth [6].

Though enough literature is present on AI but they are presented as case reports and case series in the literature. Very few studies were available with a proper study design, thus this systematic review was initiated in an attempt to assess the effectiveness of restorative treatment of AI in children.

Objectives: To assess the effectiveness of direct and indirect restorative materials in patients with amelogenesis imperfecta.

Material and Methods

Eligibility Criteria

Inclusion and exclusion criteria were fixed and the studies were screened based on the criteria mentioned below:

Inclusion criteria

1. Studies done on children with AI of any type.
2. Study designs like RCT, clinical trial, prospective studies, retrospective studies.
3. Studies which use any of the direct or indirect restorative materials for AI patients.
4. Studies in English or studies in other languages where translation to English is possible
5. Studies published from the year 1st January 2001 to 31st October 2019.

Exclusion Criteria

1. Studies wherein the review parameters are assessed without clinical examination.
2. Studies which are done on patients who have other diseases along with AI.
3. Reviews
4. Case reports
5. Case series
6. Conference proceedings
7. Letters to editor
8. Short communications
9. *In-vitro* studies

PIO from PICO can be referred as

1. **Patient Population:** Children affected with AI
2. **Intervention:** Restorative treatment (direct or indirect)
3. **Outcome:** Treatment modality, failure, success rate, longevity, patient's satisfaction

Information Sources

A search strategy was developed using keywords related to AI and restorative treatments. Data was searched through the database, PubMed and Google scholar from 1st January 2001 till 31st October 2019. Cross-references were checked for relevant articles; grey literature was also searched on treatments for AI. Hand searching of articles were done when the full texts of the relevant studies were not available through electronic database.

Search

The comprehensive data search was performed in PubMed and Google scholar. While carrying out the search through PubMed filters were put for the dates of publication as1st January 2001 till 31st October 2019. The filter on article type was set for comparative study, clinical study, clinical trial, observational study and species as humans. No language restrictions were put though studies included were in English language but studies reported in other language were also selected and then subjected to Google translation to obtain the data in English language. Studies were excluded through language only if the data cannot be translated in English. The keywords for search were decided by reviewing the literature and given in Table no.1.

Table 1: The keywords required for search

Primary keywords	Secondary keywords
Amelogenesis imperfecta (P)	• Congenital Enamel Hypoplasia • Hypoplastic type amelogenesis imperfecta • Local Hypoplastic type amelogenesis imperfect • Autosomal Recessive amelogenesis imperfect • ADHCAI • Amelogenesis Imperfecta hypomineralization Type
Children (P)	• Offspring, Adult • Children, Adult • Adolescent • Infant • Toddler • Adult Offspring • Children • Child • Preschool • Preschool Child • Children, Preschool • Preschool Children
Restorative treatment (I)	• Dental care • Restoration • Crown
Assessment (O)	• Failure • Success rate • Longevity • Life Quality • Health-Related Quality of Life • Health Related Quality of Life • HRQOL • Patient Outcome Assessments • Outcome Assessment, Patient • Patient Outcomes Assessment • Patients satisfaction • Esthetic

Table 2: The search strategy used for searching articles

Sr. No.	Search Strategy	Articles in Hit	Articles Selected
1	amelogenesis imperfecta AND children AND restoration	46	12
2	amelogenesis imperfecta AND children AND restoration NOT case report	23	11
3	restoration AND amelogenesis imperfecta AND child AND esthetic	20	8
4	restoration AND congenital enamel hypoplasia AND preschool	11	2
5	ceramic AND amelogenesis imperfecta AND children	10	4
6	restoration AND amelogenesis imperfecta AND children AND randomized controlled trial	4	4
7	amelogenesis imperfecta AND child AND dental care AND life quality	3	2
8	amelogenesis imperfecta AND crowns AND patient satisfaction	11	5
9	amelogenesis imperfecta AND child AND composite	31	7
10	Others	0	0
	Total	159	55

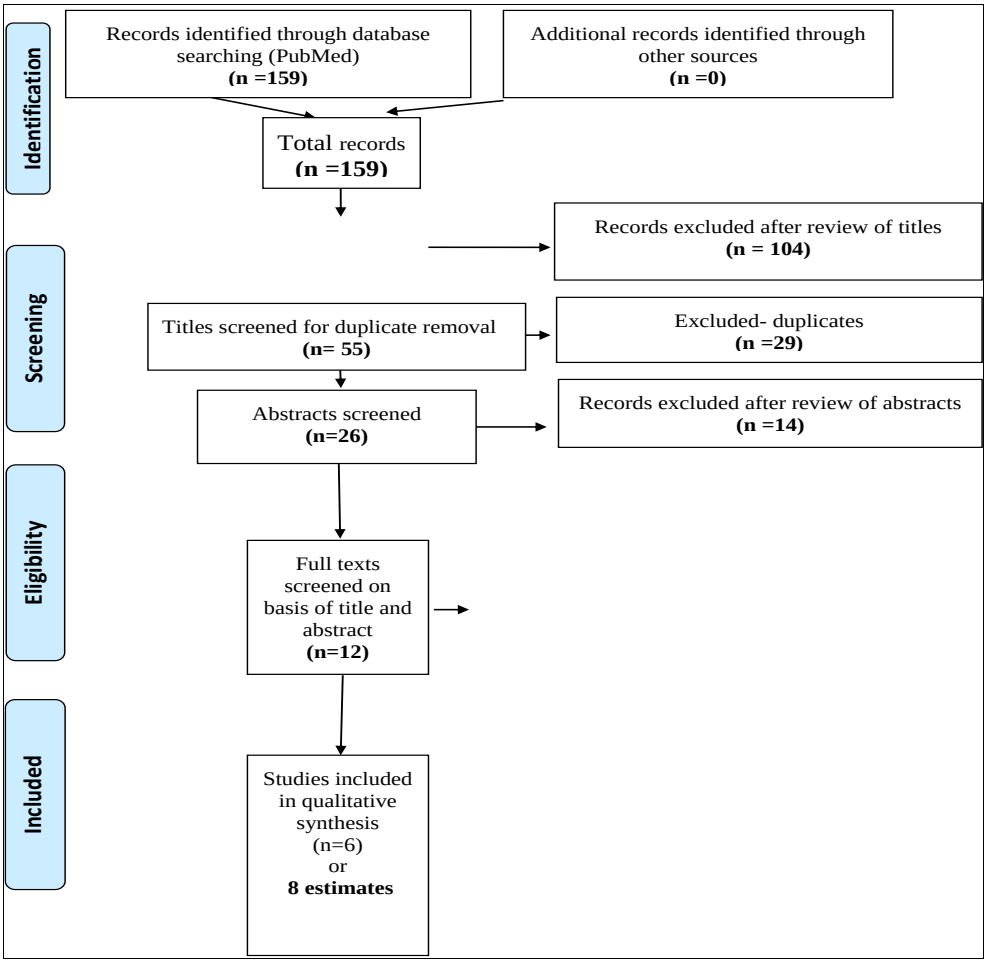
Search Strategy
The search strategy used in PubMed for searching articles is given in Table No.2. (filters set for Clinical Study, Clinical Trial, Comparative Study, Observational Study, https://www.ncbi.nlm.nih.gov/pubmed?cmd=historysearch &querykey=25 and publication dates from 1st January 2001 to 31st October 2019)

Study Selection
One review author RA independently screened the titles and abstracts obtained by search strategy and included them if they met the inclusion criteria. Later full texts of all the included studies were obtained. After that they were screened by reading the whole article and then decided if they met the inclusion criteria. Whenever there was uncertainty regarding any study to be eligible for inclusion, the problem was resolved by discussing it with the second author VB. Finally, the search yielded 55 studies to be included in the systematic review process which went further through titles, abstracts and full text review.

Data Collection Process
A standardized data extraction form was prepared in Microsoft Excel with the help of an expert. Initially 3-4 entries were made in the Excel and it was reviewed by an expert. Any disagreement between the authors was resolved by discussion. This sheet was named as pilot sheet.

Data Items
Data items included for extracting the data were: -
1. Study Id- Number given to each included study
2. Author's name- Name of the author
3. Year of publication- Year in which the study was published
4. Study design- Whether the study was clinical trial, randomized controlled trial, prospective or retrospective study
5. Defect- Which type of amelogenesis imperfecta was present in the patients
6. Extent of defect- Teeth affected with amelogenesis imperfecta, whether localised or generalized irrespective of the dentition type
7. Sample size- Sample size for that particular study (number of teeth affected or number of patients affected)
8. Treatment provided- The type of treatment provided to the patients
9. Follow-up- The time period for which the study was conducted with follow-up
10. Outcome- the outcomes of the study
- Failure- Number of teeth showing failure of the treatment provided
- Success rate- Percentage of cases with success
- Longevity- For how long did the restoration stay good without much deformity in it
- Post treatment discomfort- The immediate problem patients faced due to the treatment provided
11. Reason for failure- Explanation for the failure of the crown or restoration
12. Adverse events- Any complications which followed the treatment
13. Inference- The conclusion of the study
14. Remark- The remarks by the author (….).

Results
Study selection PRISMA Flow Chart



Discussion

Summary of Evidence

Various case reports and case series are present in the literature which explains about different treatment modalities in patients with AI. Though, they do not provide a clear idea regarding the treatment as they involve limited number of patients. Thus, in this review an attempt was made to identify studies apart from case reports and series so as to contribute to the evidence-based practice. This systematic review yielded a total of 159 articles through PubMed search strategy. No articles were identified via other sources. After screening these studies for titles, 104 studies were excluded. The remaining 55 studies were subjected for duplicate removal. A total of 29 articles were duplicates which kept coming in different search strategies. The abstract of the remaining 26 articles were read and 14 of them were excluded based on abstract. The remaining 12 articles were then read full text and a final decision was made whether to include or exclude the studies. After reading full text, 6 articles were excluded with reason: Treatment provided was of opacities, case series, case reports, same study with increased follow-up period, treatment done of cleft palate patients, data identified through interview without oral examination. Thus, a total of 6 articles were included for the evidence of the effectiveness of various treatments modalities in patients with AI. The characteristics of the articles included in this systematic review are discussed below:

Zagdwon AM *et al* [8] conducted a study which assessed two methods for the restoration of permanent molars affected by AI. It was a prospective clinical trial which was carried out on 17 patients affected by AI or severe enamel defects of first permanent molars. Two treatment modalities were SSC and other to be cast adhesive coping. A split mouth design was used so that each right or left permanent molar in both jaws was restored using either a preformed metal crown (SSC) or a cast adhesive coping (CAC). Patients were followed for up to 24 months and assessed for longevity and quality of the restorations. Among 17 patients the treatment was provided for 42 teeth and records for 42 restorations (19 SSC; 23 CAC) were present. Three restorations, one SSC at 6 months and two CAC at 2 and 19 months failed and required replacement. The study inferred that SSC was considerably cheaper to use and needed single visit, and less tooth tissue was lost in preparation and fitting and CAC was significantly more expensive but left nearly the entire tooth crown intact. Thus, the choice of the restoration depends completely on the immediate and long-term needs of each individual patient.

Vitkov *et al* [9] conducted a study which addressed the problem of AI. This study involved five patients with primary teeth affected by AI; impressions were made without previous preparation by rotary instruments. Composite crowns and veneers were manufactured and luted adhesively using the total bonding technique and low-viscosity resin composite. The preoperative oral examination revealed tooth discoloration, masticatory disturbances, hypersensitivity, and speech problems. After placement of the restorations, patients reported improvements in tooth sensitivity, articulation, and mastication. It was proved that composite crowns are quick and easy to perform, highly esthetic, and can be applied in children younger than 4 years old.

Sonmez IS *et al* [10] conducted a study to determine the effect of deproteinization on the success of composite crowns in hypocalcified AI affected permanent teeth in intraoral conditions. Since this study included AI children and a treatment to them thus, it was also included in this systematic review. A total of 32 permanent teeth in 4 healthy children with hypocalcified AI were restored with strip crowns and composite resin. Few teeth were treated with sodium hypochlorite while other were not. Clinical success was determined by USPHS modified Ryge criteria up to 36 months. The deproteinization procedure had no effect on the anatomic form of restorations. The cervical integrity of restorations showed inferior results after 36 months compared to baseline. No recurrence of caries was observed in the patients after treatment with crowns. It was inferred that the deproteinization do not have any effect on the restorations & the success rate of the restorations was good. However, composite restorations were clinically successful in children affected by hypocalcified AI in long-term follow-up.

Chen CF *et al* [11] conducted a study to assess restorative treatment outcomes in the mixed dentition of AI patients and determine the post rehabilitation oral health status and satisfaction of the patients. After identification of the patients with AI, clinical and radiographic examinations were performed on eight patients, who had 74 restorations placed in permanent incisors and molars, to allow evaluation of the integrity of the restorations and periodontal status post-treatment. Subjects completed a survey regarding esthetics, function, and sensitivity. Among the 74 restorations evaluated, seven were lost; of the remaining restorations, 31 were posterior, and 36 were anterior. Patients had reduced gingivitis, periodontitis conditions. Subject's recall of satisfaction regarding esthetics and sensitivity while brushing and eating showed a statically significant difference before and after treatment. It was concluded that during mixed dentition, teeth with AI may be restored with conventional treatment modalities. Direct restorations should be considered "interim" with multiple repairs anticipated. Post-treatment, gingival inflammation and plaque accumulation were observed.

Lundgren GP and Dahllof G *et al* [12] conducted a study to compare oral health and longevity of dental restorations in a group of young patients with AI compared to a control group. Patients included were 82 patients with AI out of which 40 were boys and 42 were girls with a age range of 6 to 25 years old. Oral examination was performed on all the included patients to assess the status of dental caries, gingivitis, previous therapy, replaced restorations, tooth sensitivity, and number of dental visits. Data was obtained from the previous record extending from 6 to 10 years before the study. It was found that an annual mean number of dental visits in the AI group were 2.9. DMFS was 8.1. The longevity of dental restorations was significantly lower in the patients with AI, with 24.7% of the AI group requiring replacement of fillings during the observation period. Patients with hypomineralized/hypomaturized AI had restorations of shorter longevity than those with hypoplastic AI. Porcelain crowns had significantly longer survival than composite resin materials in the AI patients. The study inferred that there needs a restoration which would last long in these patients and there is a great importance of establishing an early permanent-therapy plan for these patients to avoid frequent dental visits.

Lundgren GP *et al* [13] conducted a study to compare the quality and longevity of 2 crown types that is Procera and IPS Emax Press in adolescents and young adults with AI. Along with this adverse event related with the treatment were also to be documented. A total of 27 patients with a range of 11to 22 years of age with AI in need of crown therapy were included. This was a RCT using a split-mouth design. After placing 119 Procera crowns and 108 IPS Emax Press crowns following randomization, longevity, quality, adverse events, and tooth sensitivity were recorded. After 2 years of follow-up, 97% of the crowns in both crown groups had excellent or acceptable quality. Both Procera and IPS Emax Press crowns showed comparable quality in the prosthesis. Tooth sensitivity was significantly reduced after crown therapy. Adverse events such as endodontic complications occurred in 3% of crowns. It was inferred that it is possible to perform crown therapy with excellent results and without severe complications in young patients with AI.

In this systematic review only 6 articles could fit into the eligibility criteria. AI is a hereditary disorder and treatment modalities listed in the literature are many. One study was reported in the year 2003 which gave two estimates one with SSC and other with CAC. One was reported in 2006, 2009, 2013 and 2014 each. Another study reported in 2015 gave two estimates. From the studies included in this review few of them were clinical trials, few were retrospective studies few were prospective clinical trials while only one study was a RCT. The sample size ranged from unreported number of teeth in 5 patients to a maximum of 741 teeth affected by AI. Sample size was somewhat adequate in most of the studies.

Type I, II & IV AI was the defect found in almost all the studies except for two studies which did not report the same. The extent of defect in one study was restricted to only 1st permanent molars while in all other studies it was extensively present in all the teeth irrespective of dentition type. The age at which the treatment was initiated first was 3.5 years reported in one study.

The various types of treatment modalities reported were SSC, CAC, composite crowns, composite veneers, composite resin strip crowns, composite restorations and porcelain crown. The longest follow-up period reported by study conducted by Chen CF *et al* study of 100 months but it was a retrospective study. The least follow-up was present in Vitkov *et al* study of only 6 months. In the outcome; almost all the articles reported number treatments failed except for Sonmez IS *et al* study. The maximum failure number was found with Lundgren GP and Dahllof G study of 238 restorations failure. While the least was of only one failure reported by Zagdwon AM *et al*. No failure of the treatment was present treated with composite crowns and veneers in Vitkov *et al* study. The success rate was 100% in only one study by Vitkov *et al*. Overall the success rate of all the treatment modalities were in acceptable high range.

The longevity was maximum of 5 years and minimum of 6 months. The 6 months longevity was reported because the study did not further follow their patients to have a final status at 24th month. All the patients in the included studies were satisfied with their appearance and decreased sensitivity while brushing teeth and eating except for few with severe form of AI. No patient discomfort was reported in any of the study immediately after the placement of restorations in the patients. Reason for failure was reported to be inaccurate size of the crown in Zagdwon AM *et al* study. The adverse events reported by few patients were; recurrent caries, periodontitis development, dental trauma and development of apical periodontitis.

Limitation of this systematic review

1. Only PubMed database and Google scholar were searched.

Future recommendations

1. More RCT of different restorative types should be conducted.
2. A study should include large sample size.
3. The follow-up period should be long enough so as to assess the success rate

Conclusion

Within the limitations of this systematic review, it can be concluded that treatment like composite restorations, CAC, SSC, composite veneers, zirconia crowns, composite resin strip crowns, and porcelain crowns can prove to be effective in patients with AI. These treatments are highly satisfying to the patients in terms of aesthetics and sensitivity & overall improve the quality of life of patients for a long duration. The factors to be considered while opting for the type of restoration includes, the condition of the crown, severity and also the cost affordability of the patients.

References

1. Slayton RL, Warren JJ, Kanellis MJ, Levy SM, Islam M. Prevalence of enamel hypoplasia and isolated opacities in the primary dentition. Pediatr Dent,2001:23:32–6.
2. Jedeon K, De la Dure-Molla M, Brookes SJ, Loiodice S, Marciano C, Kirkham J, Canivenc-Lavier MC, Boudalia S, Berges R, Harada H, *et al*. Enamel defects reflect perinatal exposure to bisphenol A. Am J Pathol,2013:183:108–18.
3. Witkop CJ Jr. Amelogenesis imperfecta, dentinogenesis imperfecta and dentin dysplasia revisited: problems in classification. Journal of oral pathology,1988:17:547–53.
4. Crawford PJ, Aldred M, Bloch-Zupan A. Amelogenesis imperfecta. Orphanet J Rare Dis,2007:4:17.
5. Coffield KD, Phillips C, Brady M, Roberts MW, Strauss RP, Wright JT. The psychosocial impact of developmental dental defects in people with hereditary amelogenesis imperfecta. J Am Dent Assoc,2005:136:620–30.
6. Chen CF, Hu JC, Bresciani E, Peters MC, Estrella MR. Treatment considerations for patient with Amelogenesis Imperfecta: a review. Braz Dent Sci,2013:16:7–18.

7. Pires Dos Santos AP, Cabral CM, Moliterno LF, Oliveira BH. Amelogenesis imperfecta: report of a successful transitional treatment in the mixed dentition. J Dent Child,2008:75:201–6.
8. Zagdwon AM, Fayle SA, Pollard MA. A prospective clinical trial comparing preformed metal crowns and cast restorations for defective first permanent molars. Eur J Paediatr Dent,2003:4:138–42.
9. Vitkov L, Hannig M, Krautgartner WD. Restorative therapy of primary teeth severely affected by amelogenesis imperfecta. Quintessence Int,2006:37:219–24.
10. Sönmez IS, Aras S, Tunç ES, Küçükeşmen C. Clinical success of deproteinization in hypocalcified amelogenesis imperfecta. Quintessence Int,2009:40:113–8.
11. Chen CF, Hu JC, Estrella MR, Peters MC, Bresciani E. Assessment of restorative treatment of patients with amelogenesis imperfecta. Pediatr Dent,2013:35:337–42.
12. Pousette Lundgren G, Dahllöf G. Outcome of restorative treatment in young patients with amelogenesis imperfecta. a cross-sectional, retrospective study. J Dent,2014:42:1382–9.
13. Pousette Lundgren G, Morling Vestlund GI, Trulsson M, Dahllöf G. A Randomized Controlled Trial of Crown Therapy in Young Individuals with Amelogenesis Imperfecta. J Dent Res,2015:94:1041–7.