

## Dentinogenesis Imperfecta Type II in Children: A Scoping Review

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*Dentinogenesis Imperfecta type II (DI2), also known as hereditary opalescent dentin, is one of the most common genetic disorders affecting the structure of dentin, not related with osteogenesis imperfecta, which involves both primary and permanent dentitions. The purpose of this article is to perform a scoping review of the published peer-reviewed literature (1986-2017) on DI2 management in children and to outline the most relevant clinical findings extracted from this review. Forty four articles were included in the present scoping review. According to the extracted data, the following are the most important tasks to be performed in clinical pediatric dentistry: to re-establish the oral mastication, esthetics, and speech, and the development of vertical growth of alveolar bone and facial muscles; to reduce the tendency to develop caries, periapical lesions and pain; to preserve vitality, form, and size of the dentition; to avoid interfering with the eruption process of permanent teeth; to decrease the risk of tooth fractures and occlusion disturbances; to return the facial profile to a more normal appearance; and to prevent or treat possible temporomandibular joint problems. Therefore, Pediatric Dentists should bear in mind that early diagnosis and treatment, together a long-term follow-up of DI2 in children, continue to be the best approaches for achieving enhanced patient psychological well-being and, in consequence, their quality of life.*

**Keywords:** *Dentinogenesis Imperfecta Type II, Management, Children, Scoping Review.*

### INTRODUCTION

**D**entinogenesis Imperfecta (DI) is an inherited disorder that affects the dental connective tissues, mainly the organic dentin matrix, resulting in dentin structural defects both in primary and permanent dentitions.<sup>1,2</sup> The disorder was first described in the late 19<sup>th</sup> and early 20<sup>th</sup> centuries,<sup>3</sup> and Hodge *et al*<sup>4</sup> coined the name *hereditary opalescent dentin*. The term *dentinogenesis imperfecta* was introduced by Robert and Schour.<sup>5,6</sup> Genetically, DI follows an autosomal dominant Mendelian trait with nearly 100% penetration and a low mutation rate.<sup>7-9</sup> Its incidence is 1 in 6,000-8,000 individuals in the U.S., without gender predilection and affects primarily Caucasians.<sup>10-12</sup>

DI is characterized by discolored, translucent, and opalescent teeth, ranging from gray to yellow, blue, or brown.<sup>1,2</sup> In 30% of affected patients, the enamel layer is thinner than normal, hypoplastic, or hypocalcified, and poorly supported by the underlying dentin.<sup>13</sup> Therefore, under strong masticatory pressures, the enamel may fracture and break off; subsequently, the exposed dentin is severely and rapidly worn.<sup>10,14</sup> Radiographically, there are bulbous crowns, cervical constrictions, shorter roots, smaller pulp chambers, and partially or totally obliterated root canals due to continual tertiary or reparative dentin formation;<sup>13,15,16</sup> the pulp chamber and canals may be initially abnormally wide (*shell teeth* appearance), but they become obliterated or clogged progressively.<sup>6,13,15</sup> Histologically, dentinal tissue is atubular, interglobular, and severely hypomineralized, with a lower amount of odontoblasts, and pulpal inclusions are common.<sup>2,6,13,14,16</sup>

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The first DI classification was proposed by Witkop *et al*<sup>3</sup> these authors suggested that the term *hereditary opalescent dentin* be used only for cases of isolated traits, and *dentinogenesis imperfecta*, for conditions associated with osteogenesis imperfecta.<sup>3,17</sup> Shields *et al*<sup>18</sup> created the most popular classification of DI, dividing it into three basic types: (i) associated with osteogenesis imperfecta, due to a defective biosynthesis of type I collagen; (ii) associated with osteogenesis imperfecta, which is the most common form of DI, also known as *hereditary opalescent dentin*, and (iii) the *Brandywine form*, which is inhabited by a tri-racial population, and where this very rare disorder is found exclusively;<sup>1,16,19–21</sup> this later type is characterized by multiple pulp exposures in both dentitions.<sup>22</sup> Later, de La Dure-Molla *et al*<sup>21</sup> proposed a new classification of DI, as follows: *radicular dentin dysplasia* (type I of former classification); *mild/moderate forms of dentinogenesis imperfecta* (type II of the former classification), and the *severe form of dentinogenesis imperfecta* (type III of the former classification). DI types I and II are similar clinical, radiographic, and histologically; both types are caused by a mutation of the *Dentin SialoPhosphoProtein Gene (DSPP)*.<sup>9,21,23–29</sup>

The diagnosis of *dentinogenesis imperfecta* type II (DI2) is based on a detailed clinical and radiographic examination, family medical history, and genetic counseling, including the family pedigree or construction of a family tree.<sup>1,2,7,9,30,31</sup>

Currently, management of DI2 with severe dental attrition in children still represents a challenge for pediatric dentistry practitioners, taking in account the substantial loss of dental tissue and the need for functional and esthetic restorations from very early ages.<sup>32</sup> Several clinical case reports on the treatment of DI2 in children have been published in which diverse restorative techniques and materials are recommended. In this context, this scoping review was conducted to map key concepts of dental management approaches for children with *dentinogenesis imperfecta* type II available in literature, in order to identify, summarize and describe those strategies aimed to improve the oral health of this population.

## MATERIALS AND METHOD

A scoping review was conducted between October 2017 and January 2018. We implemented the Arksey and O'Malley methodological framework and recommendations for this type of reviews.<sup>33</sup>

### Research question

The present scope review intended to answer the following question research: What are the best dental management approaches for children and adolescents with *dentinogenesis imperfecta* type II in primary or young permanent teeth?

### Identifying relevant studies

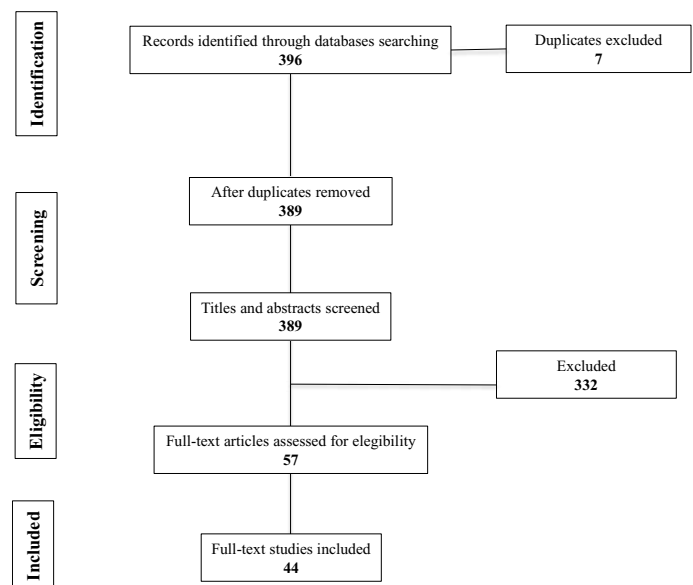
The scope review's primary objectives were: to pose a research question; to identify and retrieve relevant studies; to select those studies relevant to the research question to chart the critical data from the selected studies; and finally, to collate, summarize, and report the results from those most important articles on DI2 in children, published between January 1986 and January 2018, in order to answer the previously posed research question. These articles were accessible in three electronic databases (PubMed, Embase/Ovid, and the Cochrane Library). The search strategy was appropriately adapted for each database. The main search terms,

MeSH or free-text terms, keywords, and Boolean operators, alone or in combination, chosen for these aims included “*dentinogenesis imperfecta*”, “*dentinal dysplasia*”, “*hereditary opalescent dentin*”, “*Capdepon teeth*”, “*children*”, “*pediatric dentistry*”, “*dentistry for children*”, and “*pedodontics*”.

### Study screening and selection

Titles and abstracts derived from electronic and hand searches were carefully reviewed by two authors (ID-Z and RM-R) for selecting the most relevant of these. Different types of peer-reviewed publications, only in the English language, were screened as follows: clinical trials; observational or descriptive studies (cohort, case-control and cross-sectional studies); pilot studies; narrative reviews of the literature; *in vitro* studies; clinical case series, and case reports. A complementary hand-search in reference lists of selected articles was also performed; then, the eligible articles, in their full-text form, were obtained. The data extraction process—employing a predesigned and standardized form—was carried out by other two experienced authors (JAG-R and AJP-G) in an independent manner, and any disagreement or discrepancy was resolved by discussion and consensus with a third reviewer (MSR-R); specifically, data about DI2 diagnosis methods, oral prevention approaches, restorative/pulpal in primary and young permanent teeth and orthodontics/dentofacial orthopedics treatment modalities, and oral health maintenance, were searched for extraction. The flow chart for the whole article search process is depicted in Figure 1.

Figure 1. Flow chart for the search strategy.



## RESULTS

### Charting the data

The initial electronic database search yielded in total 396 potential articles. By title and abstract screening and after removing duplicates, 57 articles were included. These were retrieved in full-text and critically analyzed; an additional hand-search was also performed. Finally, 44 articles were included in the present scoping review for the pertinent information extraction process. The articles were classified into four categories according study design (Tables 1-4).

**Table 1. Relevant articles (Case/Case series report) included for the current scoping review (1986-2017), and their main findings.**

Author (s)-Country	Main findings
Schneidman <i>et al</i> , 1988 <sup>34</sup> -US	Two cases: a 10-year-old girl and a 13-year-old boy Both patients were treated with partial or complete overlay dentures, fabricated over retained teeth or roots
Darendeliler-Kaba <i>et al</i> , 1992 <sup>35</sup> -Switzerland	A growing child treated with overdentures placed over severe attritioned teeth
Mayordomo <i>et al</i> , 1992 <sup>36</sup> -Spain	An 11-year-old girl Dental management was a combination of restorative, prosthetic, and surgical treatments.
Raji <i>et al</i> , 1993 <sup>37</sup> - India	A three case report taken from an Indian family with DI2 Clinically and radiographic examination was performed The pedigree was traced through 4 generations No evidence was osteogenesis imperfect was found in any patient
Cehreli <i>et al</i> , 1996 <sup>38</sup> -Turkey	A 4-year-old boy with extreme attrition in primary dentition was treated with complete overdentures to preserve the clinical crowns, reestablish the vertical dimension and esthetics
Joshi <i>et al</i> , 1998 <sup>39</sup> -India	A 17-year-old girl Severely worn permanent dentition and multiple retained root stumps The patient was treated with overdentures
Tanaka <i>et al</i> , 1998 <sup>17</sup> -Japan	A 11-year-old girl The patient was treated with an overdenture to improve her occlusal height
Sapir <i>et al</i> , 2001 <sup>14</sup> -Israel	An 8-month-old girl Pedoform strip crowns filled with composite in incisors and canines Stainless steel crown for second primary molars
Huth <i>et al</i> , 2002 <sup>24</sup> -Germany	A 4-year-old boy Complete restorative treatment using composite and stainless steel crowns, under general anesthesia
Singh <i>et al</i> , 2004 <sup>40</sup> -India	A 6-year-old girl Extreme attrition of primary teeth and very narrow or obliterated pulp canals Dental procedures were aimed to pain relief and infection control Pulp treatments and overdentures
Kamboj <i>et al</i> , 2007 <sup>5</sup> -India	Two cases with 3 generation antecedents Authors suggest that severe DI2 in children can be treated in two early stages: the first one at 18 to 20 months, and the second one at 28 to 30 months
Shetty <i>et al</i> , 2007 <sup>41</sup> -India	Authors mention that DI2 dental treatment is intended to protect primary/permanent teeth from severe wear and tear Other purposes of treatment are to improve esthetics, vertical dimension, soft tissue support, and facial profile
Bouvier <i>et al</i> , 2008 <sup>32</sup> -France	A 6-year-old girl The patient was managed during 9 years, under a two-stage treated Stage 1 (primary and mixed dentitions): stainless steel crowns with subgingival preparation, and polycarbonate resin crowns Stage 2 (permanent dentition): 24 individual low-fusion ceramic-metal crowns
Delgado <i>et al</i> , 2008 <sup>31</sup> -Spain	Two cases: A 6- and a 20 year-old patients No enamel present on dental surface of primary/permanent teeth Dental treatment consisted in extractions, fissure sealants, stainless steel crowns, composite resins, and overdentures Child's esthetic appearance improved greatly
Subramaniam <i>et al</i> , 2008 <sup>6</sup> -India	An adolescent female patient exhibiting anterior gingival recession, attrition, complete enamel loss, multiple pulp exposures and periapical lesions Treatment was performed under antibiotic prophylaxis and general anesthesia, and included several extractions, root canal treatments, stainless steel and polycarbonate crowns, and removable partial dentures
Leal <i>et al</i> , 2010 <sup>19</sup> -Brazil	A 7-year-old boy Treatment: Extractions, fissure sealants, esthetic restorations, and a removable partial upper denture
Millet <i>et al</i> , 2010 <sup>42</sup> -France	A 9 year-old girl Congenitally missing of 3 permanent teeth Prosthetic treatment consisted mainly of low-fusion metal ceramic restorations 10 years of follow-up with satisfactory results
Scarel-Caminaga <i>et al</i> , 2012 <sup>10</sup> -Brazil	A 6-year-old boy Dental treatment consisted in composite resin full-coverage restorations to protect the exposed dentin, and fissure resin sealants on first permanent molars Posterior cross bite was managed with an Hawley upper removable expander

**Table 1. Relevant articles (Case/Case series report) included for the current scoping review (1986-2017), and their main findings (continued).**

Author (s)-Country	Main findings
Rafeek <i>et al</i> , 2013 <sup>43</sup> -Trinidad and Tobago	Two family members with DI2 They were treated with amalgams, composite veneers, crowns, bridges, and overdentures Authors concluded that in severe cases of severe tooth wear, the treatment of choice is full-coverage crowns. Bonded veneers are specific for improving the esthetics of anterior teeth
Surendra <i>et al</i> , 2013 <sup>44</sup> -India	A 3-year-old boy Loss of enamel and severe reduction of vertical dimension Dental treatment consisted mainly of pulpotomies in both first primary molars, and placements of stainless steel crowns
Min <i>et al</i> , 2014 <sup>45</sup> -Republic of Korea	An 11-year-old boy with multiple teeth fractures The dentin hardness was measured through the Vickers test In this patient, the dentin was almost 5 times softer than normal dentin
Khot <i>et al</i> , 2015 <sup>2</sup> -India	An 8-year-old boy Pulp obliterations in primary teeth, widened pulp spaces and cervical constriction in permanent teeth Extractions, stainless steel crowns for posterior teeth, and polycarbonate resin crowns for anterior teeth
Mori-Ubaldini <i>et al</i> , 2015 <sup>11</sup> -Brazil	An 8-year-old-boy with Class III malocclusion Problems with mastication, esthetics and speech Endodontic therapy, reconstruction of anterior teeth with direct resins
Silva-Moreira <i>et al</i> , 2015 <sup>30</sup> -Brazil	A 5-year-old boy Rehabilitation treatment was performed through acetate crowns and orthodontic bands
Akhlaghi <i>et al</i> , 2016 <sup>46</sup> -Iran	3.5 year-old boy Coronal height of teeth was reduced with exposed dentin Extractions, band-fixed space maintainers, pulpotomies, and stainless steel crowns
Beltrame <i>et al</i> , 2017 <sup>47</sup> -Brazil	1.8 year-old girl Excessive wear and fractures of enamel layer Direct and indirect composite restorations The patient was treated under midazolam/chloral hydrate sedation
Gama <i>et al</i> , 2017 <sup>48</sup> -Brazil	3.0 year-old girl Photopolymerizable composite resins The patient was followed-up over a 17-year period Third molars were surgically extracted and histopathologically analyzed by scanning electron microscopy
Ierardo <i>et al</i> , 2017 <sup>49</sup> -Italy	5.0 year-old girl Second class malocclusion, deep bite, and lower arch anterior crowding The orthodontic/orthopedic treatment consisted of <i>Nite-Guide</i> and <i>Occlus-o-Guide</i> (series G and N) until 9 years old

**Table 2. Relevant articles (In vitro) included for the current scoping review (1986-2017), and their main findings.**

Author (s)-Country	Main findings
Siar, 1986 <sup>50</sup> -Malaysia	Sample: 9 teeth affected by DI2, examined by a quantitative histological technique for coronal dentin Three basic patterns of distribution of dentin tubules were observed According to authors, this finding supports the concept of abnormal dentinogenesis in DI is attributable to a diminution or lack of normal functional odontoblasts
Lukinmaa <i>et al</i> , 1996 <sup>51</sup> -Finland and Italy	Aim: To describe the dentin structural abnormalities of permanent teeth of DI patients As a consequence of the poor quality of the organic matrix, the dentin is defectively mineralized
Kim <i>et al</i> , 2004 <sup>52</sup> -US	Defects in the human gene encoding DSPP cause inherited dentin defects, which can be associated with bilateral progressive high-frequency sensorineural hearing loss
Gallusi <i>et al</i> , 2006 <sup>16</sup> -Italy	Aim: To examine the morphology of DI-affected permanent teeth enamel, dentin and dentine-enamel junction (DEJ) The major findings were: enamel regularly mineralized, anomalies in DEJ, and dentin with absence of tubules
Lee <i>et al</i> , 2008 <sup>28</sup> -Republic of Korea	Aim: To report clinical findings and the results of mutational analysis of a Hispanic family with DI2 Authors identified a novel mutation in a splice acceptor of the DSPP in the studied family.

Author (s)-Country	Main findings
De la Dure-Molla <i>et al</i> , 2015 <sup>21</sup> -France	A new DI classification is proposed to simplify diagnosis: Radicular Dentin Dysplasia Dentinogenesis imperfecta - Mild form - Moderate form - Severe form
Li <i>et al</i> , 2017 <sup>53</sup> -China	Aim: To describe the characteristics and the genotype analyses (dentin sialoprotein and DSPP) performed in seven Chinese families affected by DI and dentin dysplasia

**Table 3. Relevant articles (*Clinical trials*) included for the current scoping review (1986-2017), and their main findings.**

Author (s)-Country	Main findings
Harley <i>et al</i> , 1993 <sup>54</sup> -UK	Sample: 20 patients aged between 7 and 18 years affected by either amelogenesis or dentinogenesis imperfecta In total, 64 adhesive cast restorations were placed without tooth preparation Full occlusion was restored in all participants
Malgrem <i>et al</i> , 2003 <sup>25</sup> -Sweden	Introduction of 2 DI scoring systems for analyzing the degree of dentin dysplastic manifestations Sample: 81 DI teeth and 20 healthy teeth Useful also for osteogenesis imperfecta
Holappa <i>et al</i> , 2006 <sup>26</sup> -Finland	A mutational analysis of dentin sialophosphoprotein (DSPP) gene causing DI, carried out in 7 Finnish affected families Through analysis of known DSPP mutations, authors suggest that DI is caused by aberration of normal splicing
Kim <i>et al</i> , 2007 <sup>27</sup> -US	According to the authors, due to significant dental attrition and loss of vertical dimension, many persons suffering DI2 exhibit abnormal mandible position and the shape of the inner ear. This can lead to hearing deficits or loss
Barron <i>et al</i> , 2008 <sup>1</sup> -UK	DI2 diagnosis is based of family history, pedigree construction, and detailed clinical examination Genetic diagnosis will be useful in the future once different mutations have been discovered
Opsahl-Vital <i>et al</i> , 2012 <sup>9</sup> -France	Authors concluded that dentin tissue constitutes a valuable tool for a better understanding of those pathological processes that affects dentin biomineralization

**Table 4. Relevant articles (*narrative reviews, new classifications of DI*) included for the current scoping review (1986-2017), and their main findings.**

Author (s)-Country	Main findings
Malgrem <i>et al</i> , 1988 <sup>55</sup> -Sweden	Authors carry out a complete clinical, radiographic and histologic description of DI2 through two branches of a 6-generation family, focusing on individual differences
Kantaputra <i>et al</i> , 2001 <sup>56</sup> -India	Author makes a brief review of DI, with especial emphasis on the disease associated syndromes
De Coster, 2012 <sup>57</sup> -Belgium	Endodontic treatment of DI teeth is very challenging because of the peculiar pulp morphology. If conventional therapy is not an option, periapical curettage and retrograde filling is a possible alternative, except in the case of teeth with short roots
De la Dure-Molla <i>et al</i> , 2015 <sup>21</sup> -France	A new DI classification is proposed to simplify diagnosis: Radicular Dentin Dysplasia Dentinogenesis imperfecta - Mild form - Moderate form - Severe form

### Collating, summarizing and reporting the results

After performing the current scoping review, a large amount of relevant clinical information from the articles included was able to be condensed. Main findings are outlined as follows:

DI2 should be clinically and radiographically differentiated from similar disorders, such as DI1, amelogenesis imperfecta, dentin dysplasia, regional odontodysplasia, tetracycline staining, irradiation or chemotherapy during root development, congenital erythropoietic porphyria, vitamin D rickets, and dental fluorosis.<sup>1,2,10,16,24,44</sup> On the other hand, DI2 has been associated with some syndromes: Ehler-Danlos syndrome type II; Goldblatt syndrome; Schimmuno-osseous dysplasia, cortical defects/wormian bones, and skeletal dysplasia with opalescent and rootless teeth.<sup>56</sup>

In the past, children with DI2 were not treated until they reached adulthood, when patients underwent all tooth extractions, which left them totally edentulous, for the fitting of a complete denture.<sup>31</sup>

The preventive approach for patients suffering from DI2 consists mainly of strict oral hygiene and dietary instructions, and topical application of fluoride varnishes.<sup>11</sup> Close/regular control and follow-up appointments for maintaining good oral health are mandatory.<sup>14,30,31,48</sup>

Severe DI2 can be initially managed in two stages in primary dentition: in an early stage, during the age of 18–20 months, and, in a second stage, around the age of 28–30 months.<sup>5,45</sup> Later, during mixed dentition, multidisciplinary management is frequently required involving other dental specialists (e.g., Prosthodontists, Orthodontists, Periodontists, and Maxillofacial Surgeons).<sup>31,36</sup> It has been recommended that dental treatment may follow three different phases: (i) Treatment of primary teeth; (ii) Provisional treatment of permanent dentition, and (iii) Definitive treatment of permanent dentition in order to re-establish function and esthetics.<sup>11</sup>

Different dental restorative modalities, single or in combination, have been suggested for both primary and young permanent teeth with severe attrition: direct and indirect restorations with composites; fixed or removable prosthetic appliances; overdentures resting on the remnants of primary teeth; stainless-steel crowns with or without esthetic front; pedoform/composite strip crowns; gold inlays; conservative adhesive cast restorations (without tooth preparation); metal-porcelain or ceramic crowns, and implant-supported restorations.<sup>2,6,11,14,30,32,36</sup> However, in severe cases of DI2, the adhesion strength of enamel and dentin is compromised.<sup>14,16,43</sup>

Overdentures re-establish esthetics, function, and vertical dimension. Also, it is possible to achieve a relatively stable occlusion that improves the patient's tolerance of future treatment procedures for worn dentition.<sup>22,35,38,39</sup> However, these may give rise to hygiene difficulties due to plaque retention gaps.<sup>24,34,36</sup>

Affected teeth tend to fracture early, due to their bulbous crowns, cervical constriction, and short and thin roots; likewise, the dentin's loose structure and reduced hardness may also be responsible for this finding.<sup>46</sup> Owing to the risk of enamel, intracoronal restorations are contraindicated.<sup>58</sup>

Pulpectomy has a very poor prognosis due to partial or complete root canal obliteration.<sup>36</sup> Thus, the majority of primary/permanent teeth with periradicular infections or radiolucencies are usually extracted;<sup>15,31,32</sup> in this regard, it should be noted that periapical radiolucencies may be visible radiographically, even in the absence of any clinically obvious pathology.<sup>58</sup> However, if pulp canals can

be found, endodontic techniques may proceed normally.<sup>15</sup> Tooth extraction is also indicated when coronal fractures occur at the gingival level or below the gum.<sup>6</sup>

Orthodontic procedures have been successfully performed in children and adolescents with diverse degrees of DI2;<sup>6</sup> for instance, elastodontic devices (e.g. *Nite-Guide*<sup>®</sup> or *Occlus-o-Guide*<sup>®</sup>) can be adequately used by these patients because they do not require considerable dental retention, necessary in other orthodontic appliances.<sup>49</sup> Further, periodontal treatments to lengthen the clinical crowns of permanent teeth can be performed with caution.<sup>31,58</sup>

In less severe cases of DI2, carbamide peroxide bleaching has been successfully employed to treat tooth discoloration.<sup>14,31</sup>

Up to the year 2012, 13 mutations had been documented in DSPP.<sup>10</sup>

### DISCUSSION

According to Arksey and Malley<sup>33</sup>, a *scoping review* comprises a type of literature review relatively recently introduced, defined as a technique conducted to 'map' rapidly the key concept from relevant literature in a specific health science research field of interest, for example in clinical pediatric dentistry. This type of review is different to systematic reviews in some aspects. First, systematic reviews focus on a well-defined question and appropriate specific study designs can be identified in advance; whilst scoping reviews tend to address broader topics, in which different study designs may be applicable. In second place, systematic review aim to provide answers to the posed question from quality assessed studies; a scoping review does not intend to address very specific questions nor, consequently, to assess the quality of selected studies. The significance of scoping reviews lies in their main purposes: (i) to examine the extent, range and nature of research activity; (ii) to determine the value of performing a systematic review; (iii) to summarize and disseminate relevant research findings; and (iv) to identify research gaps in the existing health literature.<sup>59,60</sup>

Pediatric dentistry practitioners must fully understand the pathogenesis and clinical oral cavity implications of dentinogenesis imperfecta in order to detect the disease during the primary dentition phase and to design an appropriate preventive/treatment plan.<sup>5</sup> Time of dental treatment in affected children can only be decided upon on an individual basis.<sup>11,24</sup> Of course, it is also necessary to consider the behavioral cooperation exhibited by the patient, because this is usually a complicating limiting factor during the treatment.<sup>47</sup>

The following timely preventive and corrective measures can be performed to achieve therapeutic purposes: (i) establish rapport with the patient and her/his family early in the treatment; (ii) re-establish the oral functions (e.g., mastication, esthetics, and speech) and the development of vertical growth of alveolar bone and facial muscles; (iii) reduce the tendency to develop caries, periapical lesions (e.g. dentoalveolar abscesses), and pain; (iv) preserve vitality, form, and size of the dentition; (v) avoid interfering with the eruption process of permanent teeth; (vi) decrease the risk of tooth fractures and occlusion disturbances; (vii) return the facial profile to a more normal appearance; and (viii) prevent or treat possible temporomandibular joint problems.<sup>7,14,30,31,45,55</sup> Therefore, early diagnosis and multidisciplinary treatment are required.<sup>11,31,41,36</sup> The main purpose for conducting this is to obtain a favorable prognosis, because late intervention renders treatment more complex.<sup>41</sup>

In general, the selection of restorative materials and techniques for treating DI2 depends on the patient's age and cooperation level at the beginning of the treatment, parent compliance, and the severity of dental wear.<sup>10,24,32</sup> In that clinical procedures often require an extended time in the dental chair, it is necessary to consider the use of pharmacologic sedation or general anesthesia rationally, particularly at early ages.<sup>11,24</sup>

Regarding the findings obtained here, the most important limitation of the present scoping review was that only articles in English were taken in account as part of the searching inclusion criteria. This decision can be justified based on two different studies' points of view,<sup>59,61</sup> who stated that studies demonstrating a significant effect of treatment are more likely to be published in English, are more likely to be cited by others, and produce multiple publications than studies in other languages; such studies are therefore also more likely to be identified and included in scoping or systematic reviews, which may introduce bias (*e.g. reductionism*). We are aware of this limitation; although a substantial number of articles in English were reviewed for the purposes of the present scoping review, it is possible that some papers in other languages could be missed; however, we are confident that the majority of published high quality papers were retrieved.

In summary, Pediatric Dentists should keep in mind that early diagnosis and treatment, together long-term follow-up of DI2 in children, continue to be the best approaches for achieving the enhancement of the patient's psychological well-being and, consequently, their quality of life. The selection of any treatment approach must be carefully conducted on an individual basis, according to her/his unique functional, esthetic, restorative, and occlusal needs; however, the choice principally depends on severity of tooth's structure damage. On the other hand, to obtain successful management results requires that child and parents are highly motivated to comply with all instructions on oral hygiene and nutrition.

## CONCLUSIONS

In the present scoping review, we found diverse, available dental management options as reported in the literature for this genetic disorder. However, the lack of high-level evidence from randomized and controlled clinical trials performed on affected pediatric participants was notorious. Therefore, we strongly suggest the planning and developing of such research methodologies, with the aim of testing and comparing materials and techniques for teeth with severe wearing, following the recommended statements deriving from the modern concept of Evidence-Based Pediatric Dentistry.

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