ORIGINAL RESEARCH

Hereditary opalescent dentin – A systematic review

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ABSTRACT

Introduction: Inherited disorder affecting dentin is known as dentinogenesis imperfecta. The pathognomonic features such as fracture and attrition are observed. Main etiology targets mutation in dentin sialophosphoprotein (DSPP). In order to improve aesthetics and function, early diagnosis should be done followed by appropriate management to prevent further deterioration of affected teeth. This systematic review deals with such disorder in an elaborate manner. **Materials and methods:** A detailed literature search was done pertaining to OMFP patients and their association with dentinogenesis imperfecta. Inclusion criteria include various studies done on the above based topics. **Results:** Grand total of 600 cases were identified from 300 papers published in English language literature. Of these 300, 270 were filtered narrowing down to 30 fully downloaded studies pertaining to the topic. **Conclusion:** Perspective from a dental practitioner should be done in an exceptional way in patients with OMFP in order to achieve long-term success.

Keywords: oral and maxillofacial, pathology, dentin, heredity, DSPP

INTRODUCTION

The fundamental elements of the external hard structure of a tooth, encapsulating its central portion known as the pulp, include the enamel and dentin. Cementum envelops the dentin tissue of the tooth root; however, it is categorized within the periodontal tissues (1).Dental disorders are categorized based on the impacted tissue (enamel or dentin), their nature (congenital or acquired), or the method of hereditary transmission. These conditions are further differentiated by shape and structural abnormalities, such as Amelogenesis Imperfecta or Enamel Hypoplasia, Dentinogenesis Imperfecta I, II, and III, Dentin Dysplasia type I and II, as well as various disorders like oligodontia, hypodontia, anodontia, supernumerary teeth, and others. (1–3).Dentinogenesis Imperfecta, a well-known dental condition with a longstanding history, affects dental tissues, including both tooth enamel and dentin. Dentinogenesis Imperfecta is an inherited oral condition, initially identified by Barret in 1882 (4). The term 'dentinogenesis imperfecta' was introduced by Robert and Schour in 1939 (5). It can manifest as a primary disorder or as a component of a genetic or metabolic condition. Notably, Dentinogenesis Imperfecta is frequently observed in individuals with Osteogenesis Imperfecta. The clinical presentation of this disease is highly diverse, and a comprehensive understanding of the subject facilitates early diagnosis and accurate assessment, particularly in the context of severe and rapidly progressing metabolic bone disorders. Recent advancements in the realms of biology and genetics have significantly facilitated the early detection of Dentinogenesis Imperfecta. These breakthroughs have enabled the identification of responsible genes, aiding in the determination of inheritance patterns and the recognition of potential associations with other syndromes (1). This systematic review targets on dentinogenesis imperfecta in a detailed manner.

MATERIALS AND METHODS

A comprehensive research was done. Articles from beginning to till date are considered. The literature databases included were pubmed, web of science, google scholar, scopus, medline followed by cross references. Keywords included were pathology, dentin, oral, dental, lesions. Multi journals involving oral and maxillofacial surgery, oral and maxillofacial pathology and oral and maxillofacial medicines were included. Literatures in English language which are fully available were included. The important points include publication date, author name, journal name, date of issue and keypoints.

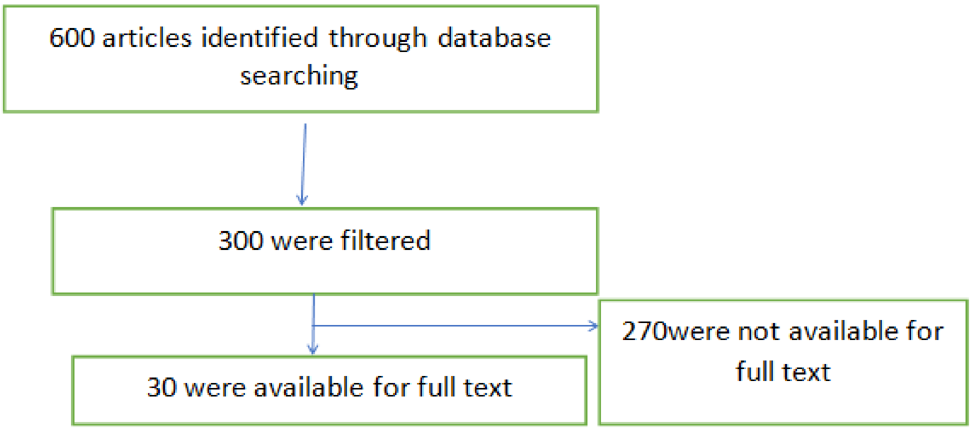

Figure 1 - Flowchart of literature search

RESULTS

Figure 2 – Dentinogenesis imperfecta (Pic. Courtesy-Dr Karthik Shunmugavelu)

The vast literature search was ended up in 30 published articles which are fully downloaded in English from various databases. The universal language of science is English. In order to avoid biasing and erroneous decisions, other languages were excluded. Duplicate articles were removed. Articles which were not able to fully download were removed. Key areas included oral and maxillofacial pathology, oral microbiology, oral and maxillofacial medicine, etc.

TABLE 1 - OVERVIEW

S.No	Literature	Author	Year	Inference
1	JRPMS.	Tsoukala E, Chochtoula G, Theodorou I, Lambrou GI.	2022	Occurrence>50%
2	Frontiers in physiology	Smith CEL, Poulter JA, Antanaviciute A, Kirkham J, Brookes SJ, Inglehearn CF, et al.	2017	Genetics to clinical via artificial intelligence
3	International journal of oral science	Lu T, Li M, Xu X, Xiong J, Huang C, Zhang X, et al.	2018	hereditary dentin disorders
4	Indian J Dent Res.	Raji MA, Vargheese NO, Grorge K.	1993	Varied colour and attrition
5	J Clin Diagn Res.	Surendra P, Shah R, N M R, Reddy VVS.	2013	histodifferentiation

6	Dent Res J (Isfahan).	Biria M, Abbas FM, Mozaffar S, Ahmadi R	2012	Osteogenesis imperfecta
7	Int J Clin Pediatr Dent.	Syriac G, Joseph E, Rupesh S, Mathew J.	2017	Gingival level
8	Acta Genet Stat Med.	Witkop CJ.	1957	hereditary
9	J Dent (Tehran).	Akhlaghi N, Eshghi AR, Mohamadpour M.	2016	DI TYPE II
10	J Dent Res.	Kerebel B, Daculsi G, Menanteau J, Kerebel LM.	1981	Defective crystallized carbonated apatite.
11	Archives of Oral Biology.	Wright JT, Gantt DG.	1985	Irregular epithelial mesenchymal interaction
12	J Adhes Dent.	Soliman S, Meyer-Marcotty P, Hahn B, Halbleib K, Krastl G.	2018	Lamination strength
13	Acta Odontol Scand.	Malmgren B, Norgren S	2002	Osteogenesis impfercta type 1 and 2
14	J Oral Sci.	Kamboj M, Chandra A.	2007	Hereditary opalescent dentin
15	Arch Oral Biol..	Shields ED, Bixler D, el-Kafrawy AM.	1973	Hereditary dentine defects
16	Am J Med Genet.	Ranta H, Lukinmaa PL, Waltimo J.	1993	Dentinogenesis imperfecta and dentin dysplasia
17	Genes (Basel).	Simmer JP, Zhang H, Moon SJH, Donnelly LAJ, Lee YL, Seymen F, et al.	2022	DSPP mutation
18	J Oral Pathology Medicine.	Witkop CJ.	1988	Dentin dysplasia
19	Eur J Hum Genet.	de La Dure-Molla M, Philippe Fournier B, Berdal A.	2015	Isolated dentinogenesis imperfecta
20	Contemp Clin Dent.	Goud A, Deshpande S.	2011	rehabilitation
21	Pediatr Dent.	-	2016	heredity
22	Pediatr Dent.	Sapir S, Shapira J.	2007	Risk assessment
23	Quintessence Int.	Hicks J, Flaitz C.	2007	In vitro
24	J Clin Pediatr Dent.	Kwok-Tung L, King NM.	2006	Mixed dentition
25	Quintessence Int.	Vitkov L, Hannig M, Krautgartner WD.	2006	therapeutic
26	J Prosthet Dent. 2004	Ozturk N, Sari Z, Ozturk B.	2004	interdisciplinary
27	ral Surg Oral Med Oral Pathol Oral Radiol Endod.	Pettiette MT, Wright JT, Trope M.	1998	endodontics
28	J Esthet Restor Dent.	• Soares C.J • Fonseca R.B • Martins L.R • Giannini M	2001	Hypoplasia
29	J Prosthet Dent.	Henke DA, Fridrich TA, Aquilino SA.	1999	Occlusal rehabilitation
30	Oral Surg Oral Med Oral Pathol Oral Radiol Endod.	O'Connell AC, Marini JC.	1999	Osteogenesis imperfecta

DISCUSSION

Dentinogenesis Imperfecta (DI) follows an autosomal dominant mode of transmission characterized by high penetrance and a low mutation rate. This condition impacts both primary and permanent dentition, with deciduous teeth typically exhibiting more severe manifestations(6). DI is identified as a localized form of mesodermal dysplasia observed during the histodifferentiation stage(5,7). Notably, it stands out as one of the prevalent autosomal dominant disorders in humans.(7) The incidence rate of DI is reported as 1 in 8000 births in the United States.(7,8)

From a clinical perspective, Dentinogenesis Imperfecta primarily manifests through distinct features, notably the discoloration of the crown. The colour spectrum observed in affected teeth varies, ranging from amber, yellow, brown, purple to even blue translucent hues. Concurrently, there is a notable occurrence of intense dental abrasion. The enamel tends to detach, exposing the underlying dentin, which may subsequently undergo sclerotic changes, resulting in a vitreous appearance. Interestingly, many patients do not report hypersensitivity, possibly due to the protective sclerotic transformation of the dentin. However, in untreated cases, the entire dentition is at risk of abrasion up to the gingival contour. Additionally, there may be instances of pronounced mobility and premature tooth loss in advanced cases (1,5–7,9).On radiographs, the teeth exhibit bulbous molars attributed to the prominent cervical stenosis. Conical roots with periapical constrictions or even the absence of roots can be noted. The density and thickness of the enamel matrix appear typical. In the initial phases of the disease, the pulp appears unusually wide, but over time, it undergoes gradual and progressive deterioration. Pulp stones are also commonly observed in many cases (1,5–7,9).Histologically, under normal conditions, the dentoenamel junction displays a scalloped pattern, facilitating the mechanical adherence of the two tissues. In Dentinogenesis Imperfecta, this characteristic scalloping is absent, resulting in a problematic connection between enamel and dentin. Consequently, there is an early loss of enamel, exposing dentin to severe and rapid abrasion. While mantle dentin may exhibit normal characteristics, atubular dentin showcases diminished calcification and a reduced number of odontoblasts. The dental tubules, in this context, appear coarse, sparse, and irregular, with an overall decreased number (1,5–7,9–11). Also noted histologically is the irregular structure of apatite crystals. The impacted dentin contains reduced levels of calcium (Ca), phosphorus (P), and magnesium, with an elevated Ca:P ratio, and the elevated levels of water and collagen in the organic composition of dentin. (12–14)\

There are two widely recognized classification systems for Dentinogenesis Imperfecta, one developed by Witkop(8) and the other by Shields(15) (Table 2)(16) .The Shields classification, implemented in 1973, comprehensively encompasses the clinical spectrum of Dentinogenesis Imperfecta (DGI) and Dentin Dysplasia (DD) by incorporating three distinct types of DGI and two types of DD. This classification system has become integral to understanding and characterizing these dental disorders in both academic dental education and clinical application.(5,7,17)

Table 2- DENTINOGENESIS IMPERFECTA

SHIELD	CLINICAL PRESENTATION	WITKOP
Dentinogenesis Imperfecta I	Osteogenesis Imperfecta with opalescent teeth	Dentinogenesis Imperfecta
Dentinogenesis Imperfecta II	Isolated Dentinogenesis Imperfecta	Hereditary Opalescent Dentin
Dentinogenesis Imperfecta III	Isolated Dentinogenesis Imperfecta	Brandywine Isolate

DENTINOGENESIS IMPERFECTA TYPE I (DGI-I)

Clinical features

Dentinogenesis Imperfecta type I (DGI-I) is a rare inherited dentin abnormality characterized by opaque dentin. With a frequency ranging from 1 in 6,000 to 1 in 8,000, it impacts both deciduous and permanent dentition.(5,7) Clinical features include the presence of abnormal dentin, affecting the structural integrity of the teeth. Additionally, DGI-I is closely associated with Osteogenesis Imperfecta (OI), establishing a significant connection between dental and skeletal abnormalities in affected individuals (1,17)

Radiographic Presentation

Radiographically, DGI-I is identified by specific features such as opaque dentin, which contributes to the diagnosis of this condition.(1,17)

Genetic Mutation

DGI-I is primarily caused by mutations in the procollagen type I COL1A1 or COL1A2 gene. These genetic alterations specifically impact procollagen type I and play a crucial role in the development of abnormal dentin observed in affected individuals.(1,5,17) The genetic connection between DGI-I and OI underscores the importance of understanding the underlying genetic mutations for accurate diagnosis and comprehensive patient care. Despite this, the significant interplay between genetic disorders affecting bones and teeth, as seen in DGI-I, is not consistently explored in genetic studies. This might be attributed to historical exclusions of teeth from the skeletal system, emphasizing the need for a more integrated approach to comprehensively address genetic disorders impacting both dental and skeletal. (1,17)

DENTINOGENESIS IMPERFECTA TYPE II (DGI-II)

Clinical Features

Dentinogenesis Imperfecta type II (DGI-II) shares similarities with DGI-I in its characteristics; the term Shields DGI-II (hereditary opalescent dentine) is employed to characterize non-syndromic autosomal dominant dentin malformations, which often mirror those found in Osteogenesis Imperfecta (OI) patients, including dental discoloration however, osteogenesis imperfecta is not a concurrent manifestation. A distinctive trait of this syndrome includes bulbous molars, accompanied by pronounced cervical stenosis(1,5,7). Shields specified that both the primary and permanent dentitions exhibit equal clinical and radiographic involvement.(17)

Radiographic Features

On radiographs, the presentation includes bulbous crowns characterized by pronounced cervical constriction, abbreviated roots, and pulp chambers that undergo faster obliteration than is typically observed.(17)

Genetic Mutation

The origin of this condition can be attributed to a genetic mutation in the DSPP gene (5,7)

DENTINOGENESIS IMPERFECTA TYPE III (DGI-III)

Clinical Features

Dentinogenesis Imperfecta Type III (DGI-III) (Brandywine isolate hereditary opalescent dentin) bears notable resemblance to its predecessors. This particular type is of rare occurrence and has exclusively been identified within the Brandywine population in Maryland, USA. The clinical characteristics are diverse, and newly emerging teeth exhibit numerous pulp exposures. (1)

Radiographic Features

Inclusion of shell teeth in the primary dentition is considered within this context, although it is not deemed a mandatory characteristic. In the secondary dentitions of individuals within the Brandywine isolate, there were observations of obliterated pulp chambers, resembling radiographic features akin to DGI-II. Similarly, the permanent teeth of children who displayed shell-like teeth in their primary dentitions also exhibited radiographic similarities to DGI-II (18).

Genetic Mutation

Shields identified DGI-III as a unique form of dentinogenesis imperfecta, possibly sharing a genetic allelic association with type II.The mutation in the DSPP gene is a causative factor for this condition, similar to the DGI-II type. The DSPP gene, situated on chromosome 4q21, encodes the principal non-collagenous protein within the dentin matrix. DSPP undergoes protease conversion into three significant proteins—dental sialoprotein, dental glycoprotein, and dental phosphoprotein. Mutations in this gene result in reduced DSPP protein levels and/or improper calcification, contributing to insufficient dentin mineralization. Alternatively, the mutated gene accumulates in odontoblasts, causing cell destruction and impacting the protein processing or transport system during the rapid production of the dentin matrix (1,5,7).de La Dure-Molla, Foruner, and Berdal (2015) have introduced a novel classification intended to replace the Shield Classification from 1973. This updated classification aims to address the limitations of its predecessor, particularly the clinical challenges posed by overlapping signs and symptoms within the sub-types. (19). In their proposed classification, the authors advocate for the collective term "Dentinogenesis imperfecta" for DSPP (dentine sialophosphoprotein) diseases, encompassing both dentinogenesis imperfecta and dentine dysplasia. The sub-types are then differentiated based on the severity of the condition as mild, moderate and severe and radicular dentin dysplasia, although there are a few exceptions:

- Shields' classification delineates Dentine Dysplasia type I as a distinctive condition characterized by exclusive involvement in root development. The recent classification introduces the term "radicular dentin dysplasia" to specifically designate this condition.
- On the other hand, Shields' Dentinogenesis Imperfecta type I is not recognized in the updated classification. The authors consider it a distinct disorder, as it is identified as a syndrome of osteogenesis imperfecta. (19)

Table 3. Former and new classification for isolated dentin rare diseases

Shield classification of isolated dentin diseases	Proposed classification of isolated dentin diseases
Dentin dysplasia type I	Radicular dentin dysplasia Dentinogenesis imperfecta
Dentin dysplasia type II	Mild form
Dentinogenesis imperfecta type II	Moderate form
Dentinogenesis imperfecta type III	Severe form

Mild Type

In primary (baby) teeth, the impact is moderate, exhibiting no severe discoloration.

For permanent (adult) teeth, either no discoloration is present, or it is minimal, often manifesting as a mild grey hue. Notably, there is minimal or no evidence of attrition (tooth wear). The crowns of the teeth may exhibit a bulbous appearance with marked constriction at the cemento-enamel junction (CEJ).(19)Radiographically, there is observable evidence of partial pulp obliteration, characterized by a distinctive "thistle-shaped appearance," as described in the literature. (19)

Moderate Type

In the moderate type, teeth exhibit a moderate degree of discoloration, presenting hues such as blue, grey, or amber opalescent shades. Increased attrition is noticeable, leading to a reduction in crown height. The crowns may assume a bulbous appearance with noticeable constriction at the cemento-enamel junction (CEJ).(19)Radiographically, the pulp is either notably reduced in size or completely obliterated. The roots display a thinner and shorter profile compared to the average, and there may be indications of periapical pathology.(19)

Severe Type

In the severe type, teeth exhibit pronounced discoloration, often appearing as a distinctive brown opalescent shade. The crowns are markedly shortened, reflecting severe attrition. Bulbous crowns with prominent constriction at the cemento-enamel junction (CEJ) are characteristic.Radiographically, a substantial pulp size is observed, accompanied by thin dentine layers, resembling what is described as "shell teeth" in the Presentation section. The roots appear thin and short, and there may be multiple periapical pathologies.(19)

Radicular Dentin Dysplasia

This subtype replaces Shields' Dentine Dysplasia Type I, wherein solely the roots of the teeth are impacted.Both primary and permanent teeth display the effects of this condition.Clinically, the teeth exhibit a normal appearance. Radiographically, there is evident root shortening, and the roots appear fused with a rounded apex.(19)

DIFFERENTIAL DIAGNOSIS

Differential diagnoses for Dentinogenesis Imperfecta, presenting similar clinical and radiographic features, encompass various conditions:(5,7,20)

- Dentin Dysplasia.
- Amelogenesis Imperfecta - hypocalcified type III, resulting in enamel loss and dentin exposure.
- Internal discolorations of dental substance, including congenital erythropoietic porphyria, rhesus incompatibility, tetracycline staining, etc.
- Conditions associated with premature tooth loss, such as hypophosphatasia, vitamin D-dependent Rickets syndrome, vitamin D-resistant Rickets syndrome, immune deficiency syndromes, etc.

TREATMENT

General Considerations and Management Principles

Effectively addressing Dentinogenesis Imperfecta (DI) involves a multifaceted approach, encompassing the prevention of severe attrition linked to enamel loss, managing the rapid wear of inadequately mineralized dentin, rehabilitating dentitions impacted by extensive wear, optimizing aesthetics, and preventing both caries and periodontal disease. The dental management strategy for DI must be tailored to the specific severity of its clinical expression.(21)Clinicians must exercise caution when treating individuals with Osteogenesis Imperfecta (OI), especially when undertaking surgical procedures or other treatments that could transmit forces to the jaws, thereby elevating the risk of bone fracture. Certain forms of protective stabilization may be contraindicated in patients with OI, emphasizing the need for careful consideration and a personalized approach to treatment planning. (21)Ensuring optimal preventive and restorative care is crucial, with aesthetics being a paramount consideration. This approach aims to maintain the patient's vertical face height when the upper and lower teeth come together. Across various types of Dentinogenesis Imperfecta (DI), the foundation of treatment remains consistent, prioritizing prevention, preservation of occlusal face height, functional maintenance, and aesthetic concerns. (1)

Preventive Measures

Timely recognition and proactive interventions are imperative for individuals with Dentinogenesis Imperfecta (DI) to mitigate the potential adverse social and functional implications of the disorder. Regular and periodic examinations play a crucial role in identifying teeth that require attention as they erupt. Rigorous oral hygiene practices, including calculus removal and the use of oral rinses, contribute to enhanced periodontal health. Furthermore, fluoride applications and the application of desensitizing agents have the potential to alleviate tooth sensitivity, offering additional preventive benefits.(21–23)

Restorative Care

Routine restorative techniques prove effective in addressing mild to moderate cases of Dentinogenesis Imperfecta (DI). These procedures are more commonly employed for permanent teeth, given that the permanent dentition is often less severely affected than the primary dentition. In instances of more severe cases characterized by substantial enamel fracturing and rapid dental wear, the preferred treatment involves full coverage restorations for both primary and permanent dentitions. The optimal success of full coverage is observed in teeth with crowns and roots that closely approximate a normal shape and size, thereby minimizing the risk of cervical fracture.(24–26)Ideally, it is preferable to accomplish the restorative stabilization of the dentition before encountering excessive wear and the subsequent loss of vertical dimension. In instances where there is a notable loss of vertical dimension, there is merit in restoring a more typical vertical dimension as part of dental rehabilitation. The use of stainless-steel crowns is advocated for primary teeth, especially when occlusal face height may be significantly compromised due to attrition or enamel erosion.(27) Cases characterized by severe loss of coronal tooth structure and vertical dimension may be evaluated as potential candidates for overdenture therapy. Overlay dentures, applied to teeth coated with fluoride-releasing glass ionomer cement, have demonstrated successful outcomes (16).Bleaching has been documented to achieve some success in lightening the colour of Dentinogenesis Imperfecta (DI) teeth. However, since the predominant cause of discoloration stems from the underlying yellow-brown dentin, relying solely on bleaching is unlikely to result in a completely normal appearance, especially in cases of significant discoloration. (21) For aesthetic considerations, full-coverage crowns or veneers (composite/porcelain) are often necessary to prevent further attrition and to mask the blue-grey discolouration.(21,28) Bonding is another option, involving the application of lighter enamel on the weakened teeth. However, common cosmetic procedures like braces and bridges are generally unsuitable for Dentinogenesis Imperfecta patients, as they may exacerbate damage.

Endodontic Considerations

Proactive measures in prevention can mitigate pulp-related pathologies, potentially facilitating less challenging endodontic procedures with improved outcomes (29).In individuals with dentinogenesis imperfecta, some may experience recurrent periapical abscesses, seemingly arising from pulpal strangulation secondary to pulpal obliteration or pulp exposure due to extensive coronal wear. The likelihood of periapical abscesses serves as a rationale for periodic radiographic assessments in individuals with DI(29). Root canal treatment for DI-affected teeth poses challenges due to pulp chamber and root canal obliteration or narrowing. Given pulpal obliteration there may be a need for apical surgery to preserve teeth affected by abscesses. The endeavour to navigate and instrument obliterated canals in DI teeth carries the risk of lateral perforation owing to the poorly mineralized dentin. (21). In instances of considerable attrition, overdentures may be prescribed to prevent further attrition of remaining teeth and maintain occlusal face height. The overarching goal is to provide comprehensive dental care that addresses both functional and aesthetic aspects, tailored to the unique challenges presented by Dentinogenesis Imperfecta.(27)

Occlusion

Dentinogenesis Imperfecta Type I often presents with Class III malocclusion, accompanied by a notable prevalence of posterior crossbites and open bites, necessitating thorough evaluation. Given the complexity of the needs in individuals affected by DI, a multidisciplinary approach is crucial for comprehensive management. (30)

CONCLUSION

Dentinogenesis Imperfecta (DI) presents a complex spectrum of dental disorders affecting enamel and dentin. With autosomal dominant inheritance, DI demonstrates diverse clinical, radiographic, and histological features, requiring a tailored approach to preventive, restorative, and endodontic care. Recent advancements in genetic understanding offer opportunities for early detection and intervention, contributing to improved patient outcomes. A multidisciplinary approach remains pivotal for addressing the intricate needs of individuals affected by DI, emphasizing the importance of comprehensive oral health management.

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