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The dental needs of children with Epidermolysis Bullosa and service delivery: a scoping review

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Abstract

Background Epidermolysis Bullosa (EB) is a genetic condition with fragility of the skin and oral mucosal lining requiring appropriate care and management by dental health professionals. The objective of this scoping review was to comprehensively examine the specialised dental needs of children with Epidermolysis Bullosa and map evidence towards the type, availability, and accessibility of specialised dental care services within various health care systems.

Methods This scoping review was conducted using the JBI Methodology framework for scoping reviews. Five databases were used to source relevant literature: MEDLINE, Embase, Dentistry & Oral Sciences Source, Scopus, and Web of Science during the period 1963–2022.

Results Thirty three published case reports were identified reporting on 45 participants encompassing the dental care and management of children diagnosed with EB aged between 0–12 years of age from an Australian and international health care context. The findings reveal the need for greater awareness amongst health professionals in the management and specialised dental care needs of children and the need for further research, and care pathways for children with EB.

Conclusion There is a dearth of evidence which examines the dental needs of children, in particular referral pathways and timely access to dental health services and professionals. Dentists play an important role in monitoring and providing individualised and specialised oral care and treatment to the child with EB. It is vital that dentists as well as the wider multidisciplinary team have knowledge and understanding of the EB condition in meeting the specialised needs and management of these children.

Keywords Epidermolysis bullosa, Dentistry, Paediatric, Specialist dental service, Health services

Introduction

Epidermolysis Bullosa (EB) is a rare inherited disease affecting the skin and mucosal membranes in response to minor trauma. The condition has thirty reported sub types across four main classifications of the disease based on the blister formations noted as: EB Simplex (EBS), Junctional EB (JEB), Dystrophic EB (DEB) and Kindler EB (KEB) [1]. The type of EB can range from mild to severe in nature impacting an estimated 500,000 people globally [1]. The condition is incurable and affects people

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from birth with chronic fragility of the skin, blistering, ulcerations, and trauma to the skin and mucosal membranes from minor injury, trauma, rubbing, friction, and heat [2, 3]. Babies born with this condition are commonly referred to as *'butterfly children'* due to the thin, fragile, and translucent nature of their skin similar to that of a delicate butterfly's wings [1]. Children with EB have been reported to experience traumatic stress reactions from not only their medical treatments but interactions with health professionals providing painful treatments [4, 5]. Similarly, continued daily EB treatments and management of their condition has also reported to impact and cause strain on the individuals, their family and those providing care [4–6].

Dependent on the type of EB, the eyes, nails, and hair can also be affected in addition to the mouth, gums, throat and esophagus, stomach, and bladder [1]. For children, blistering and trauma to the oral mucosa can impact their ability to eat and maintain healthy weight, nutrition, growth, and wound healing [7]. Of the four types of EB all patients experienced some degree of mouth ulceration. EBS is identified as having milder oral cavity ulceration [1]. However, JEB, DEB and KEB have additional health issues of tooth enamel decay, tooth decay, overcrowding or misalignment of teeth, and oesophageal blistering [1]. The sub type KEB also has additional oral cavity complications of gingivitis, tooth decay, loss of teeth and gingival enlargement (*growth of the gum around the teeth*) [8]. For the EB child general oral health care is complex with a focus on preventative care, the management of oral hygiene, dental caries, and necessary tooth extractions [5, 9]. Similarly, dental sensitivity, pain and oral care in general are areas often overlooked for children with this condition. Several authors report children as reluctant to conduct daily cleaning, thereby being non-compliant with ongoing recommended treatment/care when visiting dentists thereby increasing the incidence of ongoing dental treatment issues such as infection, teeth cavities and inflammation of the gums and overall poor oral health [10–12]. For children with EB regular in the chair dental treatment can be painful and traumatic with further trauma and complications experienced to the oral mucosa, with many children refusing treatment based on fear, pain, and previous negative dental experiences [13].

Children with EB may undergo numerous invasive procedures with their condition further compromised in the regular dental environment due to non-compliance, pain, trauma, and further complications to the oral mucosa. Dental care for EB children is often required to be undertaken in the operating room setting under general anaesthetic where specialised care and management can be fulfilled in a safe and controlled environment. Little is currently understood of the current arrangement of dental services, accessibility, and the availability of healthcare services for EB children.

Current guidelines on dental care for EB patients focus on prevention and management with a shared care approach with the multidisciplinary team providing care [8]. Referrals to specialist dental services are often required for the management of painful extractions or dental treatments which are unable to be performed in a regular dental clinic. For many children, dental treatment is best undertaken within the perioperative setting with experienced anaesthetic staff familiar with the EB condition as anaesthetic management can be hazardous with issues such as difficultly establishing an airway during intubation and trauma to the airway [14–16]. Many children with EB have had successful surgical procedures conducted under a general anaesthetic, with new techniques to manage the airway successfully intraoperatively in a controlled environment to improve their long-term oral EB rehabilitation to delimit exacerbating further oral, skin trauma and integrity during the perioperative period [17–20].

Globally, there is extensive literature of challenging and complex dental EB cases and the necessity for individualised dental care and management across the lifespan [20–24]. It is imperative that health services and schemes are available to assist patients with ongoing care requirements across the lifespan. In the Australian context, there is support by the National Disability Insurance Scheme (NDIS) for children and adults with significant physical impairment for the severe types of EB whilst those with milder forms of EB are unsupported in meeting their specific care requirements [25]. Therefore, there is a need for greater attention on the dental needs and care requirement for the EB child in line with their developmental oral health needs. Dental guidelines in managing EB patients have emphasised the need for early access to dental services with regular prevention and monitoring by a local dentist [8, 26]. In effect, the local dentist is a primary conduit for a shared care approach and referral to specialised dentistry services should sedation or general anesthesia be required to aid in the child's ongoing management and improve the quality of oral health outcomes across the lifespan [21]. Access to regular and specialised dental services may not always be readily available. This may impact how children and families who may require individualised preventative care, access treatment and care to manage their condition and their long-term oral health. Improving dental care and services for children with EB is an area often overlooked and in need of highlighting for the whole multidisciplinary health team. As treatment is often required early it is important for all health professionals to have an awareness of EB and the potential impact on the child's

developmental phases, nutrition, healthy weight, growth, wound healing, speech, and oral health. The purpose of this scoping review is to provide insight into the best evidence base of specialised dental care and management for children with EB during their pivotal developmental ages between 0–12 years and map evidence towards the type, availability, and accessibility of specialised dental care services within various health care systems from the extant literature.

Scoping review questions

The following questions guided the scoping review:

1. What are the specialised dental needs of children with EB?
2. What is the availability and accessibility of specialised dental care services currently available for children impacted with EB?

Methods

Given the rare nature of EB and the dearth of literature specifically exploring the dental care of children it was decided that a scoping review was the best approach and suitable in nature to address the research topic and questions from an international perspective. To guide the review process, the scoping review was conducted in accordance with the JBI methodology framework for scoping reviews and reported using the Preferred Reporting Items for Systematic Reviews and Meta-Analyses extension for Scoping Reviews (PRISMA-ScR) [27–30] to broadly explore and map current evidence from the extant literature.

Inclusion criteria

Participants

In line with the review questions, we included children 0–12 years of age, both male and female, of any ethnicity diagnosed with any type and form of EB (*e.g., EB Simplex (EBS), Junctional EB (JEB), Dystrophic EB (DEB) and Kindler EB (KEB*).

Concept

Included were studies which examined EB dental treatments for paediatric patients requiring specialised dental care, treatment, or specialised services from various health care settings (*e.g., dental clinic, hospital setting, hospital clinic, operating room*) inclusive of care provided by the multidisciplinary health care professionals (*e.g., dentists, dental nurse, dental surgeons & anesthetists*).

Context

Studies from any geographical location, setting which reported on children 0–12 years of age with EB requiring specialised dental care or treatment or services were considered for inclusion within this review. Studies which addressed aspects of '*dental services*', '*referral processes*' and '*management of the paediatric EB patient*' were included within this review.

Types of sources

This review considered all forms of primary studies; experimental and quasi-experimental study designs including randomized controlled trials, non-randomized controlled trials, before and after studies, interrupted time-series studies, qualitative studies, and text and opinion papers published in English language.

Exclusion criteria

The following exclusion criteria were applied during the abstract, title and full-text review stages:

- Ineligible phenomena of interest or health condition
- Conference posters
- Ineligible age population e.g. studies focused on children more than 12 years.
- Studies published in another language without an English translation were excluded due to lack of time and cost of translation.

Search strategy

The search strategy aimed to locate both published and unpublished primary studies. An initial search of MEDLINE, Embase, Dentistry & Oral Sciences Source (DOSS), Scopus, Web of Science was undertaken with a librarian to identify the relevant text words, and index terms to identify and source relevant articles on the topic during October 2022. The keyword search terms used for MEDLINE were: Exp epidermolysis bullosa OR epidermolysis bullosa.ti,ab OR EB.ti,ab OR bullous epidermolysis.ti,ab OR epidermoid bullosa.ti,ab Dental care. sh OR dental care for children.sh OR oral health.sh OR exp Surgery, Oral OR exp Oral Surgical Procedures OR exp Dentistry, Operative OR dentistry.sh OR exp "Oral and Maxillofacial Surgeons" OR exp tooth extraction OR exp dental clinics OR oral hygiene.sh OR dental*. ti,ab OR ((teeth OR tooth OR dental) adj2 (extraction* OR excision OR removal)).ti,ab OR oral health.ti,ab OR dental surg*.ti,ab OR dentist*.ti,ab OR teeth.ti,ab OR tooth.ti,ab OR oral maxillofacial.ti,ab OR ((hospital outpatient OR program*) adj3 (dental* OR oral OR dentist* OR teeth OR tooth OR extraction*)).ti,ab Exp child OR exp infant OR child*.ti,ab OR preschool*.ti,ab OR pediatric.ti,ab OR paediatric.ti,ab OR minor*.ti,ab OR infant*. ti,ab OR toddler*.ti,ab Exp Australia OR Australia*.

ti,ab. These search terms and strings were further used to develop a full search strategy, including all identified keywords and index terms, to ensure they were applied, and adapted accordingly for each included database and information source. The databases searched included JBI Evidence Synthesis, Cochrane Database of Systematic Reviews, MEDLINE, Embase, Dentistry & Oral Sciences Source (DOSS), Scopus, Web of Science. Sources of unpublished studies/gray literature were also searched including Google Scholar and Open Grey. Studies published in any language were included if also available in English. Studies were not limited by a specific date range apart from the inclusion of all published papers up until September 2022.

Study selection

Following the search, all identified citations were collated and uploaded into Endnote 20 (*Clarivate Analytics, PA, USA)* [31], with duplicates removed prior to import into the JBI System for the Unified Management, Assessment and Review of Information (JBI SUMARI) (JBI, Adelaide, Australia) [32]. Following a pilot test, titles and abstracts were screened by three independent reviewers (ZS, MJ, YS) for assessment against the inclusion criteria filtering ineligible studies and those irrelevant to the review question. Studies put forward for full text review were assessed in detail by two independent reviewers (ZS, YS) against the inclusion criteria and where consensus could not be reached a third reviewer (MJ) was consulted. Studies excluded were recorded and reported noting the reasons for exclusion.

Data extraction, analysis & presentation

Data was extracted from the papers by reviewers (SN, ZS) using a data extraction tool developed by the reviewers and checked for accuracy and completeness of information extracted by (ZS). Extracted data included specific details about the participants, concept, context, setting, study methods, and key findings relevant to the review question/s is presented (Table 1). The data collected from each of the included studies was analysed by (ZS, SN) and has been presented graphically and in tabular format with a narrative summary of the tabulated results related to the reviews objective and questions exploring the types of EB, care, and management of specialised dental services for children impacted with EB.

Results

The literature search resulted in 789 articles sourced, and after removing duplicates and removing articles that did not meet the inclusion criteria, 33 publications were considered for full-text review. After reviewing the full-text articles, data extraction was carried out for these articles. All the reported literature were either case reports or case reviews published from 1963 to 2022. The PRISMA-ScR flow diagram describes the study selection process (Fig. 1).

Population characteristics

The total number of patients reported was 45, which included 23 males and 22 females. The geographic distribution and proportion of EB cases were predominantly reported from the United States (*n = 10*) [33, 36, 38, 42, 43, 45, 46, 51, 63, 64], followed by Brazil in second place with six case reports [44, 50, 52, 55, 58, 60]. Other countries reporting on EB were India [39, 56], Iran [22, 57], Turkey [54, 59], and Taiwan [37, 47], having 2 case reports each. All the other reports were from European countries: France [35], Germany [34], Italy [40, 48], Russia [62], and the United Kingdom [49]. Australia had two publications on EB [41, 53]. The ages ranged from newborns to age 12 (Fig. 2). All the studies reported on children 0–12 years of age, except for two studies who reported on not only a child but an adult patient within their reported case reviews (*highlighted in* Table 1) [40, 61].

Oral manifestation of EB

According to the literature, there are four major manifestations of Epidermolysis bullosa: EB simplex, junctional EB, Dystrophic EB, and Kindler syndrome [65]. For this review, we found EB simplex was described in seven case reports [37, 41, 42, 47, 58, 59, 62]. The Koebner subtype [37], and herpetiformis (Dowling Meara type) [47] type were the reported subtypes for EB simplex. Dystrophic EB was the most commonly reported type of EB, having 23 case reports [33, 34, 36, 38, 22, 39, 40, 44, 46, 48–56, 60, 61, 63, 64]. The Dystrophic form has two main subtypes, dominant and recessive subtypes. The recessive type was more commonly reported (*n = 10*), with two cases of the Haliopeau-Siemens subtype [34, 40], and one of the Touraine subtype [45]. Only one case was found on the mixed EB: the Kindler subtypes [35]. There were no reported oral manifestations of Junctional EB from our literature search.

The oral manifestation can significantly decrease the quality of life. The most common intraoral features in all the reports were multiple bullae, erosions and/or vesicles on the oral mucosa, including sites such as the tongue, hard palate, gingiva, and buccal mucosa. Most patients experienced limited mouth opening or microstomia due to repeated blistering and healing, leading to scarring and contractures around the lips and mouth [34, 36, 39, 44, 46, 50, 53, 54, 61]. The absence of upper and lower frenum [39], and lingual papillae (ankyloglossia) [34, 40, 44, 52] was also observed. Few patients had

Table 1 Summary of included studies

Authors	Country	Study Design	Objectives/Aim	Type of EB	Participant Details (Number of patients)	Healthcare setting/Name of setting	Dental findings	Anaesthesia	Outcomes
Album MM, Gaisin A, Lee KW, Buck BE, Sharrar WG, Gill FM. 1977 [33]	USA	Case review	Developing a technique of management that provided maximum safety and minimum discomfort	EB dystrophica	12-year-old white boy. (1)	Children's hospital: The Children's hospital of Philadelphia	Multiple carious teeth, dental erosions, gingivitis. Recurrent bullae and erosions of the tongue and oral mucosa, and blisters over hard palate and buccal mucosa	IV sedation ketamine	Full dental extractions
Azrak B, Kaevel K, Hofmann L, Gleissner C, Willershausen B. 2006 [34]	Germany	Case review	Provide dental treatment under general anaesthesia with minimal trauma	EB dystrophic Haliopeau-Siemens type (RDEB-HS)	7-year-old boy. (1)	Dental hospital: Johannes Gutenberg University dental clinic	Ankyloglossia, microstomia, multiple carious lesions, and gingivitis	GA	Extraction of primary maxillary incisors, second molars, and primary mandibular molars. Amalgam restoration of primary maxillary first molars and mandibular second molars. Fissure sealants on remaining teeth
Blanchet I, Tardieu C, Casazza E. 2021 [35]	France	Case review longitudinal with 7 years follow up	Long-term surveillance of oral conditions and response to maintenance therapy after periodontal treatment	Kindler poikiloderma	Two brothers; 7- and 3-year-old. (2)	General hospital: Competence Centre for Rare Odontological Diseases at Timone University Hospital (Assistance Publique-Hôpitaux de Marseille)	Enamel hypoplasia, gingivitis progressing to periodontitis, carious lesions, blisters and scars in the oral mucosa. Angular cheilitis, and pigmentation of lips	LA	7-year-old: Primary teeth were extracted under local anaesthesia and orthodontic treatment. 3-year-old: Restorative treatment for carious teeth. Scaling and polishing were performed for both, with monthly monitoring

Table 1 (continued)

Authors	Country	Study Design	Objectives/Aim	Type of EB	Participant Details (Number of patients)	Healthcare setting/Name of setting	Dental findings	Anaesthesia	Outcomes
Camm JH, Gray SE, Mayes TC. 1991 [36]	USA	Case review	Dental treatment and management and monitoring disease state	EB dystrophic recessive	5-year-old girl. (1)	General hospital: Wilford Hall USAF (United States Air Force), Medical Centre, Texas	Bullae and vesicle on the tongue, oral mucosa. Microstomia, limited mouth opening, and numerous carious teeth	GA	Dental extractions under general anaesthesia, and space maintainers
Chuang LC, Hsu CL, Lin SY. 2015 [37]	Taiwan	Case review	Delivery of a fixed denture for EB patient to improve quality of life and mastication	Koebner subtype of EB simplex	3-year-old girl. (1)	General hospital: Paediatric Dentistry Department, Chang Gung Memorial Hospital, Linkou	White lesions and ulcers on buccal mucosa and gingiva. Residual roots, and caries	GA	Pulpectomy with restoration, composite restoration, dental extractions, and removable dentures. Later, a fixed denture was given using a banded appliance with two Omega loops
Endruschat AJ, Keenen DA. 1973 [38]	USA	Case review	Administration of GA, dental management, and post-operative care	EB dystrophica	5-year-old white girl. (1)	General hospital: Department of Paediatric Dentistry, Children's Hospital Medical Centre	Oral mucosal lesions, and dental caries	GA	Restoration using SSC, silver amalgam, composite restoration, and scaling later
Esfahanizade K, Mahdavi AR, Ansari G, Fallahinejad Ghajari M, Esfahanizadeh A. 2014 [22]	Iran	Case review	Dental and anaesthetic management	EB dystrophica	12-year-old girl. (1)	General hospital: Mofid Children's Hospital, Shahid Beheshti University of Medical Sciences,Tehran, Iran	Microstomia, deep caries, dentoalveolar abscess, severe crowding, delayed eruption of permanent teeth, and ankyloglossia	LA and GA	Dental extractions of permanent molars under GA. For restoration of new carious lesions, GIC and composite were used under LA

Table 1 (continued)

Authors	Country	Study Design	Objectives/Aim	Type of EB	Participant Details (Number of patients)	Healthcare setting/Name of setting	Dental findings	Anaesthesia	Outcomes
Eswara U. 2012 [39]	India	Case review	Dental management	EB dystrophica	7-year-old boy. (1)	Dental hospital: Department of Pedodontics, The Oxford Dental College, Hospital and Research Centre	Microstomia, dental caries, absence of upper and lower frenum, greyish-white appearance of buccal mucosa and palate, ankyloglossia, reduced depth of lingual sulcus, absence of labial and buccal sulcus, and deep bite	LA	Dental restoration and fluoride varnish application
Galeotti A, D'Antò V, Gentile T, et al. 2014 [40]	Italy	Case review	Evaluate the efficacy of Er: YAG laser used for treating hard dental tissue in patients with EB	EB dystrophic Haliopeau-Siemens type (RDEB-HS)	8-year-old boy and 26-year-old female. (2)	General hospital: Dentistry unit of Bambino Ges`u Children's Hospital	8-year-old—Microstomia, extensive dental caries, ankyloglossia, obliteration of vestibule, and absence of lingual papillae, blood, and fluid-filled bullae. 26-year-old—absence or minimum presence of bullae, oedematous gingival tissue, and generalized enamel hypoplasia	None	8-year-old- Laser was used to treat dental caries. 26-year-old- Laser was used to remove the damaged enamel on maxillary incisors
Goldschmied F. 1999 [41]	Australia	Case review	Management of the patient utilising Begg light Wire fixed-appliance orthodontic treatment	EB simplex	10-year-old boy. (1)	Dental clinic	Angle Class II div 1 malocclusion	None	Fixed orthodontic using Begg's appliance

Table 1 (continued)

Authors	Country	Study Design	Objectives/Aim	Type of EB	Participant Details (Number of patients)	Healthcare setting/Name of setting	Dental findings	Anaesthesia	Outcomes
Hochberg MS, Vazquez-Santiago IA, Sher M. 1992 [42]	USA	Case review	Dental treatment with minimal trauma	EB simplex	Newborn girl. (1)	General hospital: Dental department, Interfaith Medical Centre	White lesion on tongue, gingiva, and buccal mucosa, blisters and denuded areas	None	No dental treatment reported
Hubbert CH, Adams JG. 1977 [43]	USA	Case review	Dental and anaesthetic management	EB	12-year-old white girl. (1)	General hospital: LeBonheur Children's hospital	Dental caries, and blisters	GA	Dental restorations
Kummer TR, Nagano HC, Tavares SS, Santos BZ, Miranda C. 2013 [44]	Brazil	Case review	Multidisciplinary care in dental management	EB dystrophica	11-year-old girl. (1)	Dental hospital	Oral bullae, dental caries, ankyloglossia, perioral fibrosis, microstomia, and depapilated tongue leading to decreased quality of life	None	Dental restorations, root canal treatment, scaling and polishing
Lanier PA, Posnick WR, Donly KJ. 1990 [45]	USA	Case review	Dental and anaesthetic management	Touraine type epidermolysis bullosa dystrophica	3-year-old white girl. (1)	General hospital: University of Texas Dental Branch Paediatric Dental clinic	Severe dental caries, anterior open bite, perioral bullae formation, mucosal bullae, gingivitis	GA	Dental pulpotomies, SSC, composite restoration, extraction of primary incisors
Lindemeyer R, Wadenya R, Maxwell L. 2009 [46]	USA	Case review	Dental and anaesthetic management	EB dystrophica	Two siblings, 8-year-old boy and 4-year-old girl. (2)	General hospital: Paediatric Dental Clinic, Children's Hospital of Philadelphia	8-year-old- Dental caries, erosions, abscessed teeth, intraoral bullae, no vestibule, severe scarring of mouth, and microstomia, 4-year-old- multiple carious teeth, microstomia, and intraoral bullae	GA	8-year-old: Dental extractions 4-year-old- restorative treatment included SSC and extractions

Table 1 (continued)

Authors	Country	Study Design	Objectives/Aim	Type of EB	Participant Details (Number of patients)	Healthcare setting/Name of setting	Dental findings	Anaesthesia	Outcomes
Liu HH, Chen CJ, Miles DA. 1998 [47]	Taiwan	Case review	Dental management	EB simplex herpetiformis (Dowling-Meara type)	Newborn girl. (1)	General hospital	White lesions and ulcerations on the tongue, buccal mucosa, and partially erupted mandibular incisor teeth	None	No dental treatment was reported
Marini I, Vecchiet F. 2001 [48]	Italy	Case review	To evaluate the effectiveness of a sucralfate suspension in reducing both pain and the number of blisters in patients with EB	EB dystrophica	2 males and 3 females, 6 to 11 years of age. (5)	Dental hospital: Department of Oral Surgery, School of Dentistry, University of Bologna	Blisters, dental caries, and gingivitis	None	Size of the blisters and pain reduced after the application of Sucralfate ointment
Marshall BE. 1963 [49]	United Kingdom	Case review	Dental and anaesthetic management	EB dystrophica	8-year-old boy. (1)	General hospital: United Cambridge Hospital	Oral bullae with superficial sloughing	GA	Dental extractions
Mello BZ, Neto NL, Kobayashi TY. 2016 [50]	Brazil	Case review	Dental and anaesthetic management	EB dystrophic recessive	5-year-old boy. (1)	General hospital: Hospital for Rehabilitation of Craniofacial Anomalies, University of São Paulo	Blisters on oral mucosa, tongue, gingiva, severe carious lesions, and microstomia	GA	Dental extractions of primary teeth
Morgan WC. 1975 [51]	USA	Case review	Dental management with IV sedation	EB dystrophica	5-year-old Caucasian boy. (1)	Dental hospital: College of Dentistry, University of Kentucky	Mucosal bullae formation, caries, broken down primary teeth, and abscess formation	IV ketamine	Dental restoration and extractions

Table 1 (continued)

Authors	Country	Study Design	Objectives/Aim	Type of EB	Participant Details (Number of patients)	Healthcare setting/Name of setting	Dental findings	Anaesthesia	Outcomes
Oliveira TM, Sakai VT, Candido LA, Silva SM, Machado MA. 2008 [52]	Brazil	Case review	Dental and psychological intervention	EB dystrophica	16-year-old white girl and 5-year-old African American boy. (2) * *Data extracted only for child < 12 years of age*	Dental hospital: School of Dentistry, University of São Paulo	16-year-old—ankyloglossia, multiple blisters, and microstomia. 5-year-old- blisters on the tongue, gingiva, and oral mucosa and advanced white lesions	Topical LA	16-year-old Dental restoration and extractions and prosthetic appliance for maxillary incisor. 5-year-old—Fluoride application
Olsen CB, Bourke LF. 1997 [53]	Australia	Case review	Dental management and long-term management with 20-year follow-up	EB dystrophic recessive	Siblings, Caucasian boys; newborn and 4-year-old. (2)	Dental hospital: Royal Dental Hospital of Melbourne	Dental caries, dental erosions, mucosal ulceration, and microstomia	Nitrous oxide relative analgesia, GA, IV sedation with ketamine	Newborn- Multiple extractions under LA and GA. Rochette bridge to replace maxillary anterior incisors, amalgam restoration, and endodontic treatment. 4-year-old- scaling, fluoride gels
Pekiner FN, Yücelten D, Ozbayrak S, Sezen EC. 2005 [54]	Turkey	Case review	Dental presentation of EB	EB dystrophic recessive	Newborn boys. (2)	General hospital: Istanbul University Faculty of Medicine and Marmara University Faculty of Medicine	White lesions seen on tongue, gingiva, buccal mucosa, vestibular obliteration of palatal rugae, lingual papillae, microstomia, dental caries, and severe blistering,	None	No dental treatment reported

Table 1 (continued)

Authors	Country	Study Design	Objectives/Aim	Type of EB	Participant Details (Number of patients)	Healthcare setting/Name of setting	Dental findings	Anaesthesia	Outcomes
Silva LC, Cruz RA, Abou-Id LR, Brini LN, Moreira LS. 2004 [55]	Brazil	Case review	Dental management	EB dystrophic recessive	8-year-old girl 1, 10-year-old boy, 8-year-old girl 2 (3)	Dental hospital: 8-year-old girl 1—Paediatric Dentistry Department. 10-year-old boy—sought treatment at the dental school. 8-year-old girl 2—sought treatment at the dental school	8-year-old girl 1- extensive caries, generalised white spots, and root fragments. Blister formation, ankyloglossia, microstomia, and occlusal disturbance. 10-year-old boy—generalised ulcerations in his mouth. 8-year-old girl 2—extensive carious lesions, limited mouth opening, ankyloglossia	None	8-year-old girl 1- Restoration of anterior teeth with GIC, and extraction of retained roots. 10-year-old boy—restoration using GIC, extractions, endodontic treatment, and topical fluoride. 8-year-old girl 2—extractions, and scaling
Prabhu VR, Rekka P; Ramesh, Swathi S. 2011 [56]	India	Case review	Dental and anaesthetic management	EB dystrophica	10-year-old Indian girl. (1)	General hospital	Dental caries, dentoalveolar abscess, microstomia, and blisters	GA	Extraction
Sanjari K, Bayani M, Zadeh HE. 2020 [57]	Iran	Case review	Conservative dental management	EB	11-year-old boy. (1)	Dental hospital: Department of Paediatric dentistry at Arak University of medical science, dentistry faculty Arak, Iran	Smooth tongue, blisters, obliteration of buccal and lingual vestibule, and dental caries	LA	Extraction, scaling and fissure sealants
Scheidt L, Sanabe ME, Diniz MB. 2015 [58]	Brazil	Case review	Dental presentation and management of EB	EB simplex	10-year-old girl. (1)	Dental hospital	Vesicles on hard palate	None	At home care, using mouthwash and aloe vera gel
Sipahier M. 1994 [59]	Turkey	Case review	Dental management	EB simplex	9-year-old white boy. (1)	Dental hospital: Department of Paediatric Dentistry, University of Gazi	Hypoplastic and pitted teeth, gingival hyperplasia, and caries	None	Restoration with amalgam, and topical fluoride

Table 1 (continued)

Authors	Country	Study Design	Objectives/Aim	Type of EB	Participant Details (Number of patients)	Healthcare setting/Name of setting	Dental findings	Anaesthesia	Outcomes
Torres CP, Gomes-Silva JM, Mellara TS, Carvalho LP, Borsatto MC. 2011 [60]	Brazil	Case review	Dental management	EB dystrophic recessive	11-year-old Caucasian girl. (1)	Dental hospital: Ribeirão Preto Dental School, University of São Paulo, Brazil	Bulla, scarring of mouth, haemorrhagic lesion, denuded tongue, microstomia, ulcerative gingivitis, and caries	LA	Scaling, restorations, and sealants
Véliz S, Huber H, Yubero MJ, Fuentes I, Alsayer F, Krämer SM. 2020 [61]	Chile	Case series	Dental management	EB dystrophic recessive	12-year-old girl, 18-year-old woman, 10-year-old boy. (3) ** Data extracted only for child < 12 years of age*	Dental hospital	12-year-old- obliteration of the buccal sulcus, severe microstomia, oral bullae and ulcers in oral mucosa. Absence of tongue papillae and palatal rugae. 18-year-old- depapilated tongue, vestibule obliteration, severe microstomia, and frequent development of oral bullae and ulcers. 10-year-old- severe crowding with protrusion of incisors, severe microstomia, ankyloglossia, and vestibule obliteration	LA	Dental extractions
Volovikov, O., Velichko, E., Razumova, S., & Said, O. B. 2021 [62]	Russia	Case review	Orthodontic treatment	EB simplex	11-year-old girl. (1)	Dental hospital: Department of Propaedeutic of Dental Diseases, Moscow, Russia	Malocclusion	None	Intraoral optical scanning for tissue imaging

Table 1 (continued)

Authors	Country	Study Design	Objectives/Aim	Type of EB	Participant Details (Number of patients)	Healthcare setting/Name of setting	Dental findings	Anaesthesia	Outcomes
Wright JT. 1984 [63]	USA	Case review	Dental and anaesthetic management	EB dystrophic recessive	Siblings, 6-year-old boy and 4-year-old girl. (2)	General hospital: University of Alabama, School of Dentistry	6-year-old- perioral contractures, bullae around lips, enamel hypoplasia, carious lesion, and abscessed teeth. 4-year-old- bulla, enamel hypoplasia, caries, and abscessed teeth	GA	6-year-old—dental restorations with amalgam, SSC, and extractions. 4-year-old- dental extractions SSC, and restoration
Yoon RK, Ohkawa S. 2012 [64]	USA	Case review	Dental and anaesthetic management	EB dystrophica	3-year-old girl. (1)	General hospital: Children's Hospital of New York, New York City, NY	Vesicles and ulcerations. Erythematous pharynx, multiple dental caries, and dental abscesses	GA	Pulpotomies, SSC composite dental restorations, and dental extraction

EB Epidermolysis Bullosa, *GA* General Anaesthesia, *LA* Local Anaesthesia, *SSC* Stainless Steel Crown, *IV* Intravenous, *GIC* Glass ionomer cement

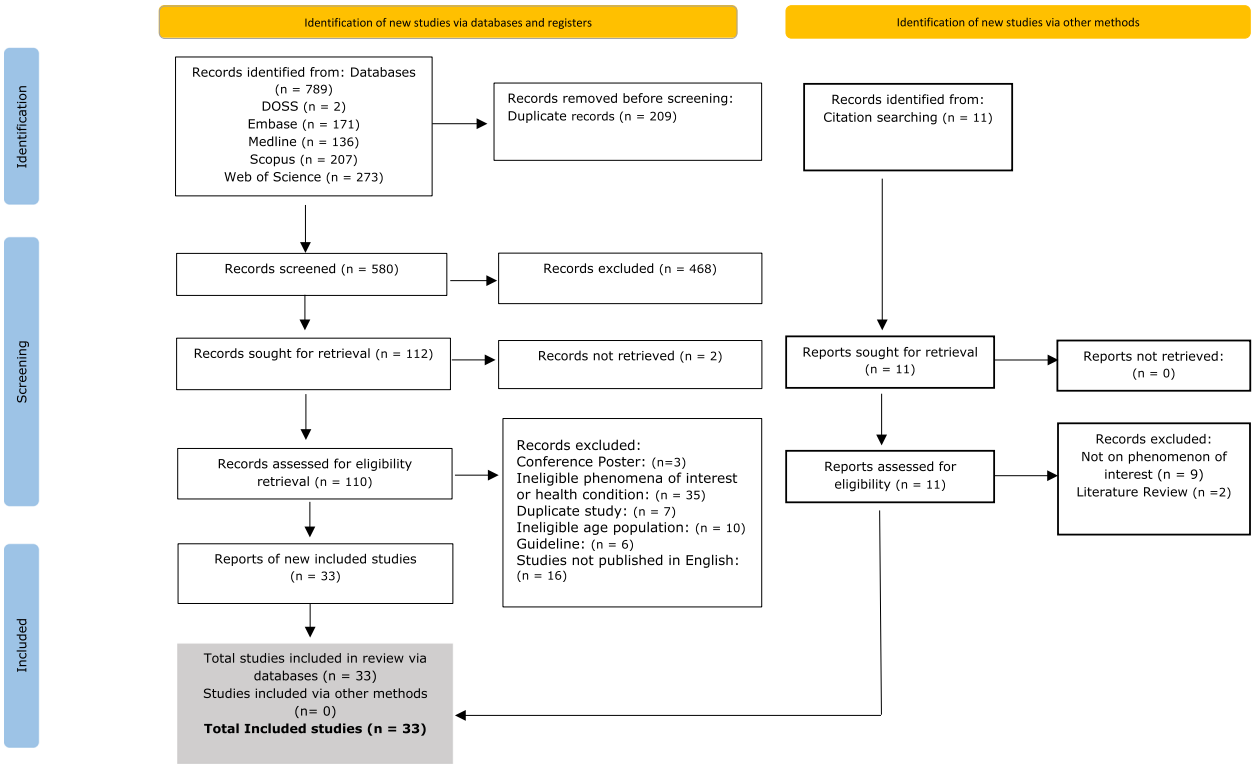

Fig. 1 Preferred Reporting Items for Systematic Reviews and Meta-Analyses extension for Scoping Reviews (PRISMA-ScR) [30]

obliteration of the buccal and lingual vestibule [39, 40, 46, 54, 61]. The tongue showed a denuded appearance without papillae [44, 57, 60], and rugae were absent from the palate [54, 61]. White lesions were observed on the tongue, gingiva, and buccal mucosa [37, 39, 47, 52, 54]. Pigmentation of lips and angular cheilitis were also reported [35].

The EB had affected both the primary and permanent dentition. Maintaining proper oral hygiene is essential for overall health, and children with EB may struggle due to pain and limited mouth opening, leading to a higher risk for dental caries and periodontal disease. The dental findings included enamel hypoplasia [35, 40] and enamel pitting [59], which progressed to carious teeth [33, 34, 36–38, 40, 43–46, 48, 50, 53, 54] and, in severe cases, led to dentoalveolar abscess formation [22, 46, 51, 56, 63, 64]. The rapid progression of caries resulted in the deterioration of teeth, leaving only remnants of root fragments [37]. There was delayed eruption of permanent teeth. Features of Class II malocclusion were also observed, showing signs of severe crowding, protrusion of incisors, and anterior open or deep bite [22, 41, 45, 61, 62]. The periodontal tissues were also affected, showing gingival inflammation or hyperplasia [59] and ulceration [60], causing gingivitis [34, 45, 48], and eventually progressing to periodontitis [35].

Healthcare treatment context & specialised treatment

The dental treatment for EB were predominantly managed in a hospital setting (*n = 18, 55%*) [33, 22, 35–38, 40, 42, 43, 45–47, 49, 50, 54, 56, 63, 64], and a dental hospital (*n = 14, 42%*), [9, 34, 39, 44, 48, 51–53, 55, 57–60, 62] with (*n* = 1, 3%) reporting orthodontic treatment provided in the dental clinic setting.63 Children requiring specialised treatment or interventions were reported within the general hospital and dental hospitals where (*n = 12*) required a general anaesthetic [34, 36–38, 43, 45, 46, 49, 50, 56, 63, 64], a local anaesthetic (*n = 5*) [9, 35, 39, 57, 60] with one case requiring a local and a general anaesthetic [22] and (*n = 2*) patients had dental care provided with IV Ketamine [33, 51], another required topical local anaesthetic [52] and one report noted a combination of treatments such as nitrous oxide, regional anaesthesia, IV sedation with ketamine [53]. Whilst (*n = 11*) were not reported to require anaesthesia for specialised treatment [40–42, 44, 47, 48, 54, 55, 58, 59, 62]. Therefore, children within this review received a variety of anaesthetic approaches to ensure optimal care and outcomes.

Dental treatment outcomes

A number of dental treatments were reported from preventative, diagnostic and restorative (Fig. 3). Dental extraction under local or general anaesthesia was

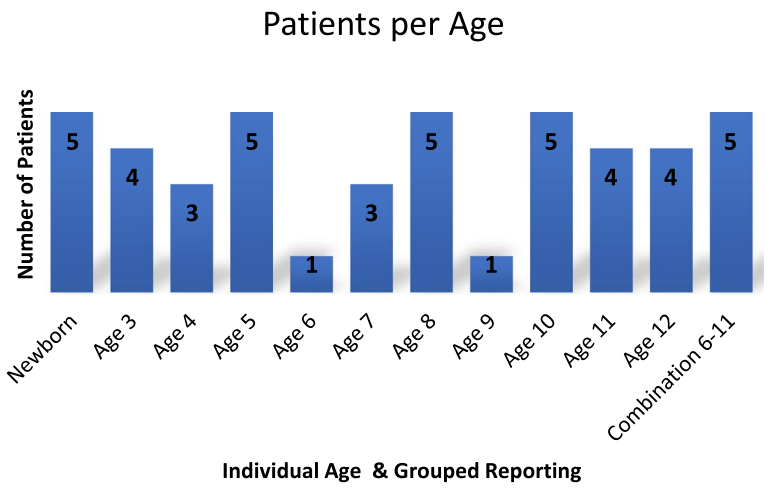

Fig. 2 Patients per age

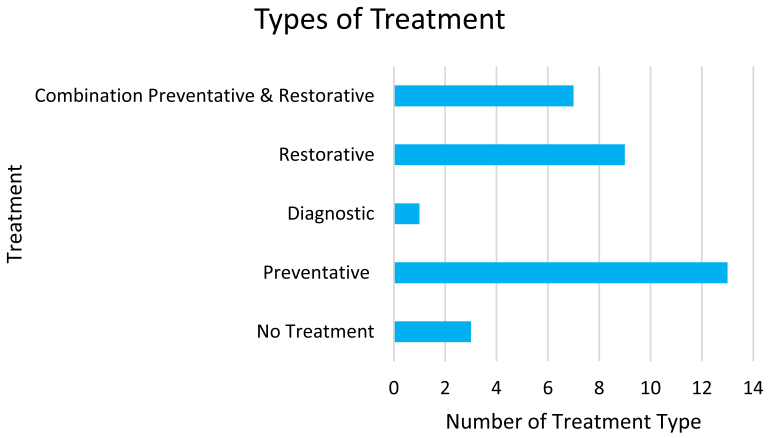

Fig. 3 Types of treatment

the most common treatment for severely decayed teeth [35, 36, 45, 46, 49, 51], or even full mouth extraction [33, 50]. For mild to moderate dental caries, the teeth were restored with GIC [22, 55] composite [38, 64], silver amalgam [34, 38, 53, 59, 63], and pulp therapy [37, 45, 64], or root canal treatment, [44, 53]was performed in severe cases. Five authors suggested using stainless steel crowns for restoring primary molars [38, 45, 46, 63, 64]. Galeotti et al. [40] reported using lasers to remove caries. In adjunctive to therapeutic and surgical treatment, oral hygiene therapy was performed [35, 38, 44, 53, 55, 57, 60], Fluoride gels [53], fluoride varnish [39, 52, 55, 59] and fissure sealants [34, 57] were used for the preventative strategies. Two authors suggested removing or fixing partial dentures to rehabilitate missing dentition for permanent dentition [37, 52], Rochette bridge, [53] and space maintainers of primary teeth [36]. Marini et al. [48] suggested home care methods such as topical application of Sucralfate on the blisters, and Scheidt et al. 47 recommended using aloe vera gel. Three authors performed removal and fixed orthodontic treatment for treating malocclusion [35, 41, 62].

Discussion

This scoping review focussed on studies primarily on children with EB exploring their dental needs and specialised treatments received. Due to the rarity of this disease and limited focus on the dental care needs of children, this scoping review comprehensively maps the evidence highlighting the complex dental and specialised care needs of forty five case reviews of children with EB informing this scoping review.

Individualised care & follow up

The need for individualised care was emphasised throughout all the case reviews presented. A number of

authors reported the need for dentists to provide continuous dental examinations from birth throughout the lifespan to monitor, recognise and address dental issues as early as possible. Therefore, predominant care in the early stages is focussed on preventative measures commencing from birth [45]. The study by Camm et al. [36] recommends triannual dental examinations, whilst other authors reported follow up monthly [35, 44] and every six months [22].

Dental compliance & health professional trust

This review has highlighted the complexity of dental care for children and the need for routine care to manage dental symptoms prior to the eruption of their first tooth and ongoing follow up care to manage their overall oral health development milestones. As recognised within this review, children requiring dental care may undergo numerous invasive procedures as a result of various dental ailments where they were unable to be managed in the regular dental environment due to complexity of care, non-compliance, pain, trauma, and further complications to their oral mucosa.

Dental compliance for any child can be difficult even more so for the child with EB where daily dental preventative treatment as simple as brushing their teeth can cause painful intraoral blistering with limited mouth opening [36, 56]. Preventative care is also related to parental knowledge, understanding of diet impacting oral health and compliance in monitoring dental hygience at home. The study by Eswara [39] highlighted this aspect reporting the experience of parents avoiding brushing their child's teeth up until aged seven years of age to avoid pain and not to cause further intra oral blistering. Therefore, parents play a pivotal role in the oral health of their child, encouraging regular oral hygiene, the use of soft toothbrushes, puree diets and supplements as required [52]. This is further supported by Torres et al. [60] who recommends diet counselling as a preventative measure to reduce potential oral health issues.

The need for timely access to dental services was also identified amongst the case reviews. The earlier study by Hochberg et al. [42] also confirmed that many patients were not brought to the dentist until they required actual care to resolve a dental issue. This was the case for a child who although from birth was diagnosed with EB was not seen by a dentist until 11 years of age until he was flagged by the dermatologist as requiring urgent dental treatment [53]. Whilst other children were seen from birth and followed through for alternate specialised care such as orthodontic treatment [41] or new innovative treatments such as sucralfate for pain and blisters [48].

For children with EB developing trust in health professionals is important particularly when undergoing painful procedures. As such for the EB child requiring specialised care, meeting new dentists, oral surgeons, and other health professionals, as well as visiting new places such as an operating room or an outpatient clinic can be a difficult and traumatic experience. Interestingly, to build trust and continuity of care with patients a few of the dentists within this review reported providing ongoing dental care across the lifespan for the child [35, 54]. This may not always be possible with specialised care and referral required elsewhere dependant on the needs of the child and compliance with treatment. This was the case when earlier authors reported issues with limited cooperation by some children with dental therapy in the chair, [33, 58] and the preference for dental therapy to involve procedures under anaesthesia [35, 36, 45, 46, 49, 51].

Specialised care & treatment in the operating room

Several authors (dentists) have discussed the need to monitor and minimise trauma to their patients when providing any form of treatment to delimit fragility of the oral tissue causing blistering [33, 48, 58] with less invasive procedures producing the best effects [40], as well as a focus on overall safety and patient benefits of procedures in the operating setting [46, 50]. Several of the case reviews reported the need for uncooperative children with EB requiring dental treatment in the operating room setting where specialised care and management could be fulfilled in a safe and controlled environment [45, 54, 63]. The risks of care under general anaesthesia was reported as primarily inflicting only minor trauma to the airway, and minor post operative complications as well as general trauma to the skin when inserting intravenous lines as well as the use of various tapes [36]. The type of treatments for EB primarily in the operating setting were reported as dental extractions and this continues to be the main surgery type. This was confirmed by Hubbert and Adams [43] earlier report noting dental and reconstructive surgery of the hands and fingers as prominent surgeries for EB within the operating room setting. Overall, access to specialised care and treatment via surgical intervention was effective in managing the child's condition with minimal trauma and pain experienced.

Other minor forms of treatment were provided in the hospital or dental clinics with clinicians preferring treatment in outpatient settings to decrease the risk of patients developing secondary infections [42]. An important aspect of EB is the need to monitor oral infections and blistering on a regular basis. Authors Yoon and Ohkawa [64], recommend the use of topical antibiotics and oral antiseptics to assist in resolving secondary infections. Whilst Hochberg et al. [42] reported the use of antibiotics pre and post dental treatment.

Dental services, individualised care, continuity of care & referral processes

Little is currently understood of the current arrangements or dental care pathways for patients with EB. As evidenced with the reports each patient has had a unique journey through the health system in receiving care at varied ages. There is a dearth of information on the nature of dental services, accessibility, and the availability of healthcare services for EB children nor how the team provides initial referral in amongst the multidisciplinary team. From the case reviews examined both internationally and within the Australian context the process of referral is an unexplored phenomenon. The case review by Lindemeyer et al. [46] reported the need for an international referral from Saudi Arabia of two siblings aged four and eight years of age for treatment in the United States encompassing anaesthetic management during surgery.

Many authors emphasise the need for patients diagnosed with EB necessitating a comprehensive dental care plan in conjunction with a conservative dental treatment plan, with a multidisciplinary care team approach to improve the quality of life for these children [44, 57, 58]. Surprisingly none of the case reviews explored and outlined aspects of referral processes or care pathways for these children. Several authors however do emphasise the need for managing the complex conditions of each patient, accessibility to care in some reports, the decision making processes in providing the best available care via intraoperative management, and safe and effective treatments across the lifespan [9, 60, 62–64]. Continuity of care is an area which requires further exploration as some children had not seen a dentist for some time, and this certainly can restrict timely care towards correcting oral health issues.

Across the studies, accessibility to dental care was not reported as a dominant issue although there are differences and perhaps disparities in health services and accessibility to dental care services globally. Interestingly from an Australian context only two papers were identified exploring the dental management of EB patients [41, 53]. A report outlining data from the EB national registry on the distribution of EB patients described a large number of people residing predominantly outside the major metropolitan areas with many living in rural and remote regions with limited access to health professionals and treatment [66].

Implications for practice & research

Many health professionals are unfamiliar with EB as a condition as well the complexity in managing and treating patients. Several of the studies reviewed commented on the need for the multidisciplinary team to work together in providing comprehensive care to EB patients. This multidisciplinary team involves, paediatricians, geneticists, dermatologists, gastroenterologist, paediatric dentists, oral surgeons, anaesthetists, mental health teams, dieticians, physiotherapists, speech, and language therapists across various settings [9]. Interestingly, nurses play a pivotal role in providing care within the hospital, the perioperative setting and community setting for EB patients however there is little literature on the specific aspects of oral and dental care provided for these patients by nurses. From a dentistry perspective, care can be complex and challenging. It is important to raise awareness of the dental needs of children with EB in amongst the multidisciplinary team to ensure early referral, management, and specialised treatment. Dentists also need to have an awareness and understanding of the EB condition, treatment and provide appropriate and timely referral to enhance the patients' oral outcomes and quality of life across the lifespan [6, 44, 57, 58]. Whilst there are international practice guidelines [10], further research on the efficacy of services and accessibility to specialised dental services is an area worth exploring further to establish care pathways and accessible services for children from birth across their lifespan.

Limitations of the review

It is acknowledged that this review was limited based on the focus of children and the absence of research articles which met the review inclusion criteria. Unfortunately, there were limited research papers with a focus on EB and dentistry limited to children zero to 12 years of age from an international context. Similarly, this review excluded papers not published in English which can be regarded as a limitation of this scoping review. Another significant limitations to this review is the number of case reviews and reports included within the review which limit the generalisability of these papers as they are predominantly case specific. On the other hand, a strength of this review was the ability to focus solely on children and capture the data as reported via the individual case reviews from an International perspective. Overall, this review, provides an insight towards the type of care provided to children, the context of this care, the treatment received and treatment outcomes as well as the types of specialised dental care services accessed across the different health care systems as well as highlighting the importance of continuity of care and best practice towards optimal oral health.

Conclusion

EB is a condition which can affect the quality of life for children with this condition. The overall findings confirm that children with EB require ongoing dental monitoring and specialised care. Therefore, as identified through

the case reports most of the children with this condition from newborn with ongoing needs and care requirements across their lifespan. The scoping review provides an insight into the need for further research. Greater attention is required on the dental needs of children, in particular referral and timely access to dental health professionals and services. The review raises awareness of EB, and the importance of health professionals and dentists working together to meet the specialised dental care needs of these children to ensure they thrive and have a quality of life.

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Conflicts of interest
The authors declare that there are no potential sources of conflict of interest towards this project.

Authors' contributions
ZS conceived and designed the scoping review, search strategy and project administration. ZS, MJ & YS contributed to article screening. SN and ZS contributed to data extraction and analysis and write up of results. ZS contributed to the write up of the manuscript and development of figures with SN, MJ & YS providing editorial review and approval of the final manuscript. All authors read and approved the final manuscript for submission.

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Data availability
Data is provided within the manuscript.

Availability of data and materials
All data generated or analysed during this study are included in this published manuscript and its supplementary files.

Declarations

Ethics approval and consent to participate
Not applicable.

Consent for publication
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Competing interests
The authors declare no competing interests.

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