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A fixed denture for a child with epidermolysis bullosa simplex

ABSTRACT

Aim *To report the caries treatment and delivery of a fixed denture for a 3-year-old girl with epidermolysis bullosa simplex (EBS).*

Case Report *EBS is manifested on the skin or mucous membranes where skin separation is easily induced by trauma. Full-mouth rehabilitation under in-patient general anaesthesia was performed to the patient in conjunction with proper pre- and postoperative care. A fixed denture was fabricated and installed to replace the extracted teeth without later causing irritation on the mucosa. The prosthesis restored aesthetics and provided comfort without imposing the burden of compliance on the patient.*

Conclusion *Aided by meticulous pre- and postoperative care and oral hygiene reinforcement, comprehensive dental treatment coupled with fixed denture delivery can greatly improve the life quality and aesthetics for children with EBS.*

Keywords *Denture; Epidermolysis bullosa simplex; Paediatric dentistry.*

Introduction

Epidermolysis bullosa (EB) is either hereditary or acquired and characterised by bullae and blisters at the site of trauma [Fine et al., 1999]. Based on the location of tissue cleavage caused by the mechanical trauma to the skin, EB can be classified into four subtypes: EB Simplex (EBS), Junctional EB (JEB), Dystrophic EB (DEB) and Mixed EB, or Kindler Syndrome (KS) [Fine et al., 2008]. In the EBS and JEB forms, blisters form within the epidermis and the lamina lucida (a component of the basement membrane), respectively. DEB is caused by scarring which is deeper in the layer of lamina densa or the upper dermis. A recent addition to the EB family, KS (where the blister formation is mixed) is characterised by trauma-induced blistering, photosensitivity, poikiloderma and skin atrophy [Lai-Cheong et al., 2009]. EB can be further divided into major and minor subtypes based on clinical and laboratory findings [Fine et al., 2008; Wright, 2010].

EB is a systemic disease that can affect multiple organs. Besides the most common symptoms of blisters and bullae on the skin, EB may be implicated in anaemia and growth retardation or manifested in the gastrointestinal tract, the genitourinary tract, the respiratory tract and the oral cavity [Nandi and Howard, 2010; Haynes, 2010]. In the case of oral complications, mucosal vesicles, bullae and abnormal teeth (such as pitted or hypoplastic enamel) may be found in EB patients [Brooks et al., 2008; Liversidge et al., 2005]. For more serious cases, oral and perioral scarring due to repetitive blistering often lead to microstomia and ankyloglossia [Brooks et al., 2008; Liversidge, 2005]. All these conditions create difficulty for dental management. Abnormal oral soft tissues and enamel structure as well as deformities of fingers may also lead to poor oral hygiene and the development of advanced caries and periodontal diseases [Brooks et al., 2008; Wright et al., 1989]. A functional dentition facilitates efficient mastication and proper nutrition intake and, in turn, helps sustaining the life quality of EB patients [Krämer, 2010; Carrol et al., 1983; Haynes, 2010], while reducing the probabilities of mucosal damage in the oral cavity and the gastrointestinal tract. Therefore, dental therapy should be an integral part of the holistic care of EB patients, especially for those suffering the severe types [Krämer, 2010; Wright, 1984].

Case report

The patient was a 3-year and 9-month-old girl with an onset of epidermolysis bullosa simplex at birth at the Chiayi Christian Hospital in Chiayi ì (Taiwan). The genetic analysis confirmed the diagnosis of the Koebner subtype of EBS where a point mutation within the chromosome 12 caused the abnormality of keratin 5 [Fine et al., 2008]. She suffered an esophageal stricture and gastroesophageal reflux disease (GERD) and was given the nasogastric tube for feeding during infancy. She usually ate soft diet and swallowed very slowly. Her parents were reluctant to brush her teeth for fear of causing pain due to the lesions in her mouth. According to the mother, the patient showed tooth erosion and fragility after the eruption. She received several pulp treatments and restorations at local clinics in Chiayi and Taoyuan. However, given her complicated dental history and fragile skin condition, the family dentists suggested that she seek comprehensive treatment at a medical center, which initiated her visit to the department of paediatric dentistry at the Chang-

FIG. 1 Multiple blisters, bullae and crusts on the patient's neck, extremities and trunk. Palmo-plantar hyperkeratosis, erosions and contractures of fingers (due to repeated scarring) were found.

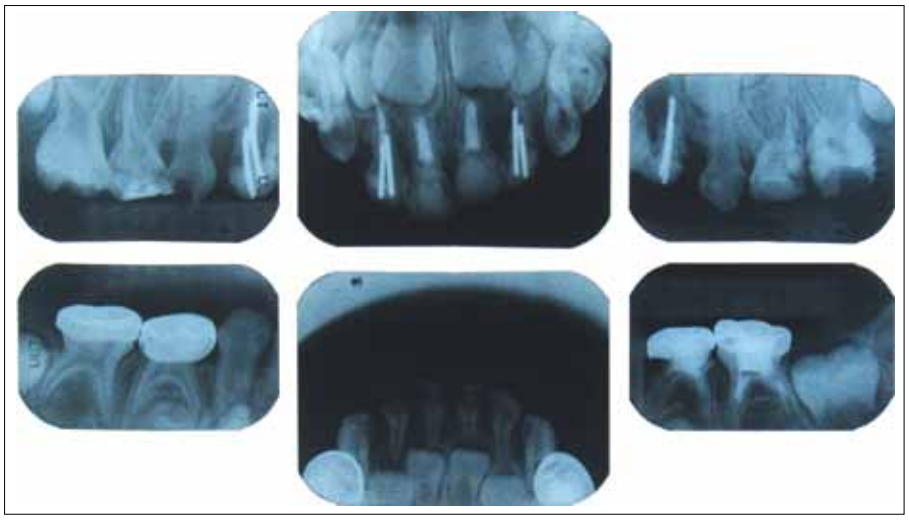

FIG. 2 Preoperative findings on periapical radiographs: teeth D, E, F and G: residual roots; tooth I: unrestorable caries; teeth A, B, C, H, J, N, O, P and Q: caries; teeth L and K: ill-fitted SSCs post pulp treatment; tooth T: an ill-fitted SSC.

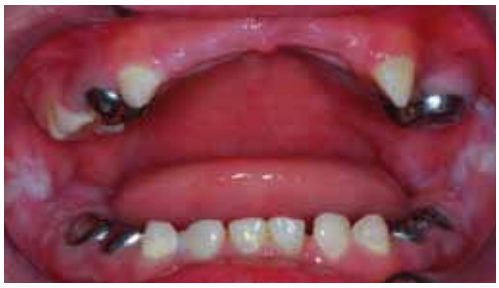

FIG. 3 No severe blisters or ulceration one week after the operation, with only mild white lesions on the bilateral buccal mucosa.

Gung Memorial Hospital in Linkou (Taiwan).

Based on the clinical examination, she suffered delayed growth due to long-term malnutrition, with a body height of 102 cm (50-75%) and a body weight of 12 kg (<3%). Multiple blisters, bullae and crusts were scattered on her neck, extremities and trunk. Palmoplantar hyperkeratosis and erosions as well as contractures of fingers (due to repetitive scarring) were also found (Fig. 1). Multiple painful lesions had rendered her gait unstable. Oral examination showed several white lesions and ulcers of various sizes on buccal mucosa and gingiva. Caries and decalcification were seen on teeth A, B, C, H, J, N, O, P and Q, and residual roots or unrestorable caries on teeth D, E, F, G and I. Teeth L and K had undergone pulp treatments with suboptimal restorations. Tooth T was poorly fitted with a stainless steel crown (SSC) (Fig. 2).

Paediatric consultation with a multidisciplinary team of geneticists, dermatologists and gastroenterologists was ordered. It was deemed necessary by the team to perform comprehensive dental treatment under general anaesthesia. A written consent form was obtained from the parent and the patient was later admitted through the paediatric department. Following antibiotics prophylaxis, preoperative complete blood count (CBC) and tests were obtained. Extra silicone and cotton padding were used and nasotracheal intubation with a flexible endoscope (for minimal trauma) was also performed. Nostril and perioral lubricant was applied for mucosal management. The following dental treatments were then carried out: pulpectomy with SSCs on teeth B and J; pulpectomy with composite restoration on tooth C; pulpectomy with strip crowns on teeth O, P and Q; composite restorations on teeth A, H and N; repeated SSCs on teeth L, K and T; and extraction of teeth D, E, F, G and I. To minimise potential mucosal trauma, the recovery period was extended before extubation. Sedative medication was also prescribed. The patient stayed in the paediatric ward for three more days before being discharged. One week after the operation, the patient showed no severe blisters or ulceration except mild white lesions on bilateral buccal mucosa (Fig. 3). Four weeks later, an impression was made for aesthetic pediatric denture, as requested by the mother. Given the patient's susceptibility to caries, we fabricated and delivered a removable denture. One month later, she complained of discomfort from the denture. We suspected that the poor retention was due to the short clinical crown of the abutment teeth and decided to change the denture appliance from the removable type to the fixed type. The fixed denture was a banded appliance fixed to the upper primary second molars and connected by a 0.040 stainless steel wire with two Omega loops, one C-clasp, four artificial primary incisors and one first primary molar (Fig. 4). Diet counseling and instructions of oral hygiene were provided to the mother. Fluoride gel, fluoride supplements and chlorhexidine were also recommended to maintain the oral hygiene and to reduce further risk of caries. With the fixed denture, the patient did not experience severe mucosa abrasion, blisters, or ulceration of soft tissue beneath the upper denture base.

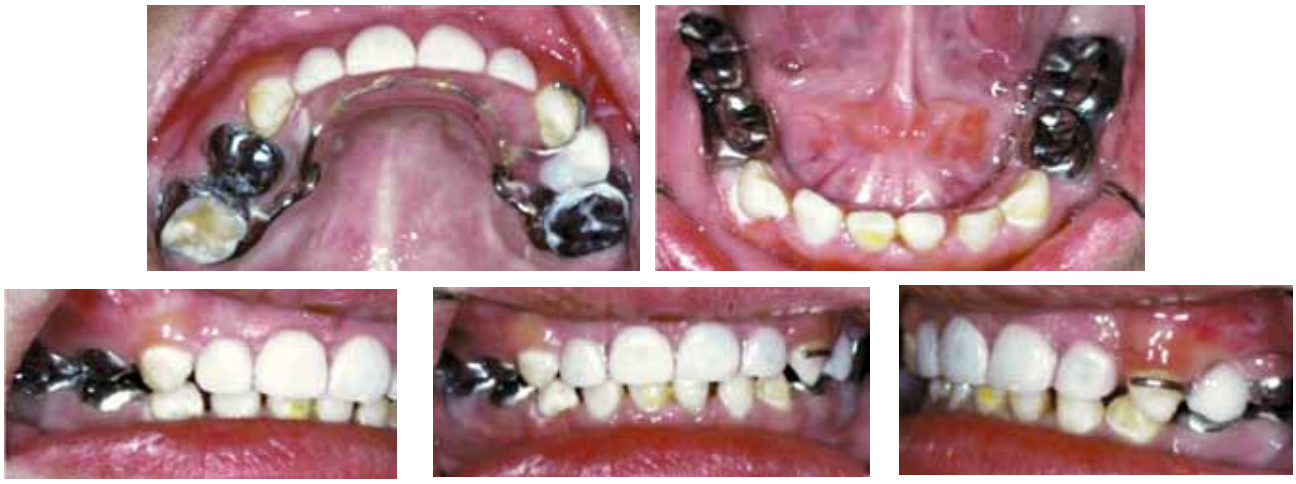

FIG. 4 The fixed denture fitted well on the patient. The denture was a banded appliance on teeth A and J, connected by a 0.040 stainless steel wire with two Omega loops, one C-clasp, four artificial primary incisors and one first primary molar.

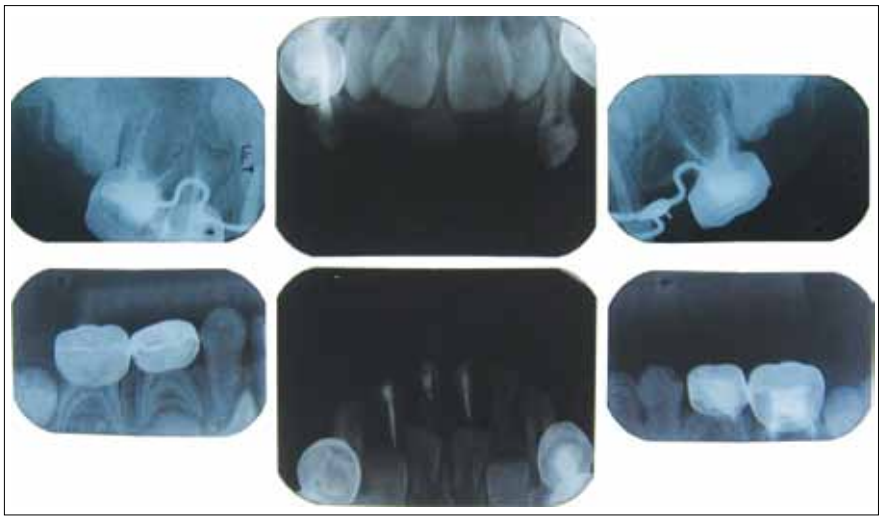

FIG. 5 Post-treatment periapical radiographs.

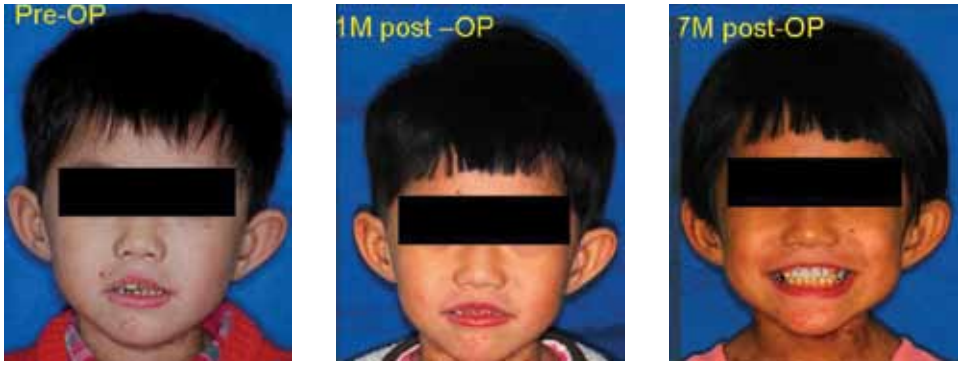

FIG. 6 Aesthetics was improved after the installation of the fixed denture.

According to her mother, the patient was able to have more solid food and her pronunciation also improved. She showed more confidence at school and proudly displayed her "new teeth" all the time. Nine months after the initial operation, however, she complained of aching pain from tooth A. We uncovered secondary caries with irreversible pulpitis on teeth A and H and performed pulpectomy treatment (Fig. 5). Her condition was stabilised and had her returned for follow-ups every three months (Fig. 6). The fixed denture was eventually removed when the patient was 6 years and 5 months (2 years and 8 months after the operation) as her permanent maxillary incisors were ready to erupt.

Discussion

EBS is the mildest form of EB, although there remain no effective therapies for EB currently. There are three common subtypes of EBS: Weber-Cockayne (localised), Koebner (generalised), and Dowling-Meara (herpetiformis) [Fine et al., 2008; Wright, 2010; Pekiner et al., 2005]. The Koebner subtype is characterised by generalised blisters and bullae and often leaves no scars once the bullae heal. Palmoplantar hyperkeratosis and erosions are common. Oral manifestations of EBS include blistering and ulceration of mucosa without enamel hypoplasia [Wright, 2010; Pekiner et al., 2005]. Typically, oral soft tissue lesions heal

without scarring, although scarring can occur with the more severe subtype such as Dowling-Meara [Wright, 2010]. Dentition and the salivary function among EBS patients tend to be normal [Wright, 1993b]. The limited oral soft tissue lesions and generally normal tooth structure among EBS patients, thus, might explain the similar prevalence of dental caries among such patients when compared to the normal populations [Wright, 2010]. However, our patient had suffered GERD and secondary osteoporosis due to malnutrition. Her teeth were eroded by gastric acid and became fragile because of mineral deficiency. Furthermore, the patient and the caregiver were reluctant to brush her teeth (which resulted in poor oral hygiene) for fear of causing pain due to the lesions in her mouth. Her predominantly soft diet and slow swallowing might have also caused the high caries rate. Out of concern over such risk, we fabricated a removable denture at first to replace the extracted incisors. Unfortunately, she complained of discomfort with the denture, which, we suspected, was triggered by the poor retention from the short clinical crown of abutment teeth and the friction from the large acrylic denture base. We then changed the denture to a fixed type, which turned out to be more comfortable with less friction and less direct contact with the ridge and mucosa. It also proved more compliance-friendly for the patient. Yet, the fixed denture did require regular cleaning and recementation every 3 months. In order to improve oral hygiene and reduce caries risk, alcohol free oral rinse, neutral fluoride gel, and fluoride supplements were prescribed to the patient [Wright et al., 1993a; Wright et al., 1994]. In conjunction with comprehensive dental treatment, a fixed-type denture greatly improved this patient's nutrient intake and esthetics. When we observed the swelling near the premaxillary alveolar bone, a sign of eruption of permanent upper central incisors, we removed the fixed denture lest the denture should restrict the premaxillary growth. One option was to use the resin-bonded denture fabricated by Dr. Orsi, instead, which had a nonrigid connector for normal premaxillary development [Orsi et al., 1999]. However, our patient had lost four primary incisors and one primary molar. A resin-bonded denture would not be suitable for her due to the long span of pontic teeth and the poor condition of the abutment teeth. This case also supports the best practice of general anaesthesia when operating on EB patients with severe soft tissue involvement [Wright, 1990a; Wright, 1990b]. Preoperative preparation should include wound care, aspiration prophylaxis, and evaluating the medication taken by the patient at the time. During general anaesthesia, atraumatic techniques should be considered, such as using the smallest endotracheal or nasotracheal tubes with lubricant. Stylet and laryngoscope can be helpful for restricted airways during intubation [Iohom and Lyons, 2001]. The patient's eyes should be lubricated, instead of being taped. Specialised dressings and tapes (Mepiform® or Mepitel®, Mölnlycke Health Care, Gothenburg, Sweden) are recommended to reduce frictions which the patient may experience from their application and removal [Nandi and Howard, 2010]. Suction needs to be carried out gently and under direct vision. Preoperative and postoperative antibiotics may be prescribed to prevent infections of the bullae. If regional anaesthesia is needed, the skin or mucosa needs to be protected and the anaesthetic solution should be injected deeply and slowly to avoid tissue separation and blister formation [Nandi and Howard, 2010; Iohom and Lyons, 2001].

Conclusion

Dental therapy should be an integral part of the holistic care of EB patients in the presence of EB's extracutaneous manifestations in the oral cavity. Aided by meticulous pre- and postoperative care and oral hygiene reinforcement, comprehensive dental treatment coupled with fixed denture delivery has greatly improved the life quality and aesthetics for this young patient with EBS during a 3-year follow-up.

References

› Brooks JK, Bare LC, Davidson J et al. Junctional epidermolysis bullosa associated with hypoplastic enamel and pervasive failure of tooth eruption: Oral rehabilitation with use of an overdenture. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2008; 105:e24-8.
› Carrol DL, Stephan MJ, Hays GL. Epidermolysis bullosa: review and report of case. J Am Dent Assoc 1983;107:749–51.
› Fine JD, Eady RA, Bauer EA et al. The classification of inherited epidermolysis bullosa (EB): Report of the Third International Consensus Meeting on Diagnosis and Classification of EB. J Am Acad Dermatol 2008;58:931-50.
› Fine JD, Johnson LB, Suchindran C, et al. The epidemiology of inherited EB: findings within American, Canadian, and European study populations. In: Fine JD, Bauer EA, McGuire J, et al, editors. Epidermolysis bullosa: clinical, epidemiologic, and laboratory advances, and the findings of the National Epidermolysis Bullosa Registry. Baltimore, MD: Johns Hopkins University Press; 1999. pp.101–13.
› Haynes L. Nutrition for Children with Epidermolysis Bullosa. Dermatol Clin 2010;28:289–301.
› Iohom G, Lyons B. Anaesthesia for children with epidermolysis bullosa: a review of 20 years' experience. Eur J Anaesthesiol 2001;18:745–54.
› Krämer SM. Oral Care and Dental Management for Patients with Epidermolysis Bullosa. Dermatol Clin 2010; 28:303-9.
› Lai-Cheong JE, Tanaka A, Hawche G et al. Kindler syndrome: a focal adhesion genodermatosis. Br J Dermatol 2009;160:233-42.
› Liversidge HM, Kosmidou A, Hector MP, Roberts GJ. Epidermolysis bullosa and dental developmental age. Int J Paediatr Dent 2005;15:335–41.
› Nandi R, Howard R. Anesthesia and Epidermolysis Bullosa. Dermatol Clin 2010;28:319–24.
› Orsi IA, Faria JFR, Bolsoni I, Freitas AC, Gatti P. The use of resin-bonded denture to replace primary incisors: case report. Pediatr Dent 1999;21:64–7.
› Pekiner FN, Yücelten D, Özbayrak S et al. Oral-clinical findings and management of epidermolysis bullosa. J Clin Pediatr Dent 2005;30:59-66.
› Wright J, Capps J, Fine JD, Johnson L. Dental caries variation in the different epidermolysis bullosa disease. J Dent Res 1989; 68:416.
› Wright JT. Epidermolysis bullosa: dental and anesthetic management of two cases. Oral Surg Oral Med Oral Pathol 1984;57:155–7.
› Wright JT. Comprehensive dental care and general anesthetic management of hereditary epidermolysis bullosa. Oral Surg Oral Med Oral Pathol 1990a; 70: 573-8.
› Wright JT. Epidermolysis bullosa: dental and anesthetic considerations-case report. Pediatr Dent 1990b;12:246–9.
› Wright JT. Oral Manifestations in the Epidermolysis Bullosa Spectrum. Dermatol Clin 2010;28:159-64.
› Wright JT, Fine JD, Johnson L. Hereditary epidermolysis bullosa: oral manifestations and dental management. Pediatr Dent 1993a;15:242–8.
› Wright JT, Fine JD, Johnson LB. Developmental defects of enamel in humans with hereditary epidermolysis bullosa. Arch Oral Biol 1993b;38:945–55.
› Wright JT, Fine JD, Johnson L. Dental caries risk in hereditary epidermolysis bullosa. Pediatr Dent 1994;16:427–32.