



## Oral health status in children with inherited dystrophic *epidermolysis bullosa*

### Stanje oralnog zdravlja dece obolele od nasledne distrofičke bulozne epidermolize

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#### Abstract

**Background/Aim.** *Epidermolysis bullosa* is a group of rare, genetic connective tissue diseases that cause blisters in the skin and mucosal membranes. The aim of this study was to evaluate the oral health status of patients with *epidermolysis bullosa dystrophica* and level of knowledge and opinion of parents about the implementation of preventive measures and quality of dental care of affected children. **Methods.** This study included a group of 17 patients from Serbia suffering from dystrophic epidermolysis bullosa and matched control group. Dental caries status was assessed using the Klein-Palmer index. Oral hygiene status was verified with oral hygiene indices, simplified plaque index, and calculus index as described by Green and Vermillion. The gingiva was assessed as healthy or inflamed (gingivitis) on the basis of any changes in color, shape, size and surface texture. The condition of oral mucosa has been registered on the basis of inspection of the lips, tongue, a floor of the mouth, mouth vestibule and palate. The level of knowledge and the impressions of parents about the application of preventive measures were investigated through two

questionnaires specifically designed for this study. **Results.** In both dentitions, there was the highest percentage of caries teeth. In primary dentition average value of the modified plaque index was  $1.4 \pm 1.14$  and modified calculus was  $0.7 \pm 1$ . On permanent teeth average plaque index was  $2 \pm 0.4$ , and average calculus  $1.6 \pm 0.6$ . Statistically, significant higher values were found in permanent dentition in percentage distribution of decayed, missing, filled teeth and also for plaque and calculus indices between affected children and the control group. Most common findings on mucosa were microstomia (76.5%) and ankyloglossia (88.2%). **Conclusion.** The absence of protocol between the treating physician and the dentist and not sufficiently informed parents are leading to inadequate dental care. The implementation of preventive measures is of most importance to decrease the risk of severe complications that are difficult to be managed.

#### Keywords:

**epidermolysis bullosa; child; oral health; preventive dentistry; surveys and questionnaires; dental caries; dental plaque index; periodontal diseases.**

#### Apstrakt

**Uvod/Cilj.** Bulozna epidermoliza je grupa retkih, genetski predisponiranih bolesti vezivnog tkiva koja se karakteriše formiranjem bula (mehurova) u koži i mukoznim membranama. Cilj rada bio je da se ispita stanje oralnog zdravlja dece obolele od distrofične bulozne epidermolize, kao i nivo znanja i mišljenje roditelja o primeni preventivnih mera i kvalitetu stomatološke zaštite obolele dece. **Metode.** U studiju je bilo uključeno 17 bolesnika sa distrofičnom buloznom epidermolizom iz Srbije i odgovarajuća kontrolna grupa. Stanje zuba ustanovljeno je Klein Palmerovim indeksom karijesnih, ekstrahovanih plombiranih zuba – KEP indeksom. Stanje oralne higijene analizirano je indeksom oralne higijene (pojednostavljeni plak indeks i indeks zubnog kamenca po Green-Vermillionu). Stanje gingive je na

osnovu inspekcije ocenjivano kao zdrava gingiva ili gingivitis, u zavisnosti od toga da li su joj bili promenjeni boja, oblik, veličina ili struktura. Stanje oralne sluzokože evidentirano je na osnovu inspekcije usana, jezika, poda usne duplje, vestibuluma, plika i frenuluma, kao i mekog i tvrdog nepca. Nivo znanja i utisci roditelja o primeni preventivnih mera i kvalitetu stomatološke zaštite dece obolele od bulozne epidermolize, ispitivani su kroz dva upitnika specijalno dizajnirana za potrebe ove studije. **Rezultati.** U obe denticije bilo je najviše karijesno obolelih zuba. Za mlečnu denticiju prosečan modifikovani plak indeks iznosio je  $1,4 \pm 1,14$ , a modifikovani kalkulus indeks  $0,7 \pm 1$ . U stalnoj denticiji vrednosti prosečnog plak indeksa i kalkulus indeksa redom su bile  $2 \pm 0,4$  i  $1,6 \pm 0,6$ . Utvrđena je statistički značajna razlika u procentualnoj zastupljenosti karijesnih, ekstrahovanih i plombiranih zuba stalne denticije između

dece obolele od bulozne epidermolize i kontrolne grupe, kao i za vrednosti plak i kalkulus indeksa. Najčešće promene oralne sluzokože bile su mikrostomija (76,5%) i ankiloglosija (88,2%). **Zaključak.** Odsustvo protokolarne saradnje između ordinirajućeg lekara i stomatologa, kao i nedovoljna informisanost roditelja, glavni su razlozi lošeg stanja oralnog zdravlja. Primena preventivnih mera je od najvećeg značaja

kako bi se sprečio nastanak komplikacija koje je teško sanirati kod ove grupe bolesnika.

#### Ključne reči:

**epidermoliza, bulozna; deca; usta, zdravlje; stomatologija, preventivna; ankete i upitnici; zub, karijes; zub, indeks plaka; periodontalne bolesti.**

## Introduction

*Epidermolysis bullosa (EB) hereditaria* is a multisystem disease that is characterized by extreme fragility of the skin and mucous membranes, on which blisters and erosions appear spontaneously or following minor trauma<sup>1</sup>. It is a rare disease, with a frequency of 1 : 17,000, and is transmitted through autosomal dominant and recessive inheritance<sup>2,3</sup>. Inherited EB is classified into four major types: simplex (EBS), junctional (JEB), dystrophic (DEB) and Kindler Syndrome<sup>4-7</sup>. DEB has two subtypes: dominant DEB (DDEB) and recessive DEB (RDEB). The worst type of EB is RDEB with the frequency of 1 : 1,000,000. Healing in patients with DEB is followed by scarring<sup>4</sup>. Diagnosis is made after birth clinically using accompanying laboratory analyses<sup>8</sup>.

There are numerous bodily systems and oral manifestations. Besides blisters and erosions on skin and mucosa, other extracutaneous involvements can be found on the gastrointestinal and in the genitourinary tract as well as within the cardiovascular and musculoskeletal system. The most difficult complication is squamous cell cancer<sup>6,9</sup>.

Common oral symptoms include blisters and erosions on mucosa, microstomia, ankyloglossia, loss of the vestibular space, absence of lingual papillae and palatal folds, enamel hypoplasia and early appearance of caries on both deciduous and permanent teeth<sup>1,6,10-12</sup>. Squamous cell carcinoma found on intraoral sites, mostly developing where tissue has experienced chronic ulcerations and repeated epithelialization. The carcinomas grow rapidly and give off metastases very early<sup>13</sup>.

The aim of this study was to determine the oral health status of DDEB patients by performing the following: dental caries assessments, oral hygiene assessments and gingival health assessments. Further assessments were made regarding the level of knowledge and opinions of parents about the implementation of preventive measures and also on the quality of dental care of EB children.

## Methods

This study included a group of 17 patients from Serbia (9 males and 8 females) suffering from DEB, aged 1-21. Two of these patients were adults with DDEB and the remaining 15 were children with RDEB. All patients were examined at the Clinic for Pediatric and Preventive Dentistry, Faculty of Dental Medicine, the University of Belgrade. Healthy controls were correspondingly matched individually by sex and age ( $\pm 6$  months) in consideration of the fact that this is a rare disease with severe symptoms and in order to obtain results that were as precise as possible.

An oral health status examination was conducted using artificial lighting, dental mirrors, and probes in accordance with criteria listed by the World Health Organization (WHO)<sup>14</sup>.

Dental caries statuses were assessed using the Klein-Palmer index for both primary (dmft<sup>1</sup>) and permanent dentition (DMFT)<sup>15</sup>. The condition of oral mucosa was evaluated on the basis of an inspection of the lips, tongue, a floor of the mouth, oral vestibule and the frenulum as well as the soft and hard palates. The mouth opening capacity was measured as the distance between the lower and upper lips vermilion lines when the patient opens the mouth as widely as possible<sup>15</sup>. The gingiva was assessed as healthy or inflamed (gingivitis) on the basis of any changes in color, shape, size and surface texture.

For the oral hygiene status in cases of primary dentition, a modified index was implemented (dental plaque index and calculus index)<sup>16</sup>. The presence of plaque and calculus was evaluated for the buccal surfaces of six representative teeth (the four second primary molars, the upper right central incisor and the lower left central incisor). For permanent dentition, a simplified plaque index and calculus index as described by Green and Vermillion<sup>17</sup> was applied (Oral Hygiene Index – OHI-S).

The level of knowledge and the impressions of parents about preventive measures in dentistry and the quality of dental care of EB children were investigated through two questionnaires specifically designed for this study (Tables 1 and 2).

Descriptions of categorical data were done using absolute and relative numbers (percentages), and for numerical data by arithmetic means and standard deviation. Frequencies were compared using the  $\chi^2$  test (if numerical conditions were not met by Fisher exact test) whilst numerical data comparison was made with the *t*-test and the Mann-Whitney U test (with and without normal distribution respectively). All applied statistical methods were considered significant if  $p \leq 0.05$ . Statistical analyses were done in IBM SPSS Statistics version 21.0 software (IBM, USA).

## Results

### Oral health status

#### Dental caries status

Among the twelve children with deciduous dentition, ten (83.3%) had affected teeth and two children (16.7%) had all caries-free teeth. From a total of 165 teeth, 95 (57.6%) were healthy and those listed with caries or as extractions or

<sup>1</sup>The decayed, missing, filled teeth index (DMFT) when written in lowercase letters-dmft, is a variation that is applied to the primary dentition.

Table 1

Questionnaire 1	
1. What type of EB has your child?	a. simplex      b. junctional      c. dominant dystrophic      d. recessive dystrophic      e. Kindler syndrome
2. After the diagnosis of EB in child is set did your treating physician recommended you to seek advice from dentist on the prevention of oral disease?	a. yes      b. no
3. What are the reasons of previous visits to the dentist (you can select more than one answer):	a. to get the advice on how to prevent and maintain oral health of my child      b. regular check-ups      c. only when the problem appears (caries, pain, swelling)
4. Does your child have regular dentist?	a. yes      b. no
5. If you can choose, who would be your choice to work with your child:	a. dentist who works in private practice      b. dentist who works in nearest community dental center      c. dentist who is trained to work with children with EB
6. During your visits to the dentist has it happened that dentist didn't dare to carry out interventions due to insufficient knowledge and experience in this area?	a. yes, dentists did not dare to carry out interventions      b. no

**EB – epidermolysis bullosa.**

Table 2

Questionnaire 2	
1. Regular check-ups are:	a. every 3 months      b. every 6 months      c. once a year      d. when problem appears
2. Dentist has informed and trained me how to prevent and maintain oral health of my child:	a. yes      b. no
3. Maintaining daily oral hygiene child uses (it can be more than one answer):	a. toothbrush and toothpaste      b. dental floss      c. oral rinses      d. all mentioned above
4. Child brushes teeth:	a. once a day      b. two times a day      c. more than two times a day      d. not every day
5. Does child brush teeth in appropriate times (in the morning before breakfast and in the evening before bedtime)?	a. yes      b. no
6. When unable to brush their teeth after a meal, do they rinse their mouth with water?	a. yes      b. no
7. Head of child's toothbrush is:	a. small      b. standard      c. I do not know
8. Hardness of child's toothbrush is:	a. soft      b. medium      c. hard      d. I do not know
9. Do you use special toothbrushes for the surfaces you can not reach?	a. yes      b. no
10. When do you replace child's old toothbrush with new one?	a. every 2-3 months      b. every 6 months      c. once a year      d. when bristles do not look nice
11. Child's toothpaste contains fluoride?	a. yes      b. no
12. What oral rinse uses your child?	a. rinse with florides      b. rinse with Chlorhexidine      c. herbal rinses      d. we do not use rinses
13. Did dentist professionally apply highly concentrated fluorides during previous dental visits?	a. yes      b. no
14. Do you use products for dental plaque identification at home?	a. yes      b. no
15. Do you know that there are medications and supplements that do not contain sugar?	a. yes      b. no
16. How many meals your child has during the day?	a. 3 meals      b. 4 meals      c. 5 and more meals
17. Each meal lasts:	a. 10 min      b. 20-30 min      c. more than 30 min
18. The food that child eats is:	a. solid but not sharp (chips, popcorn, etc.)      b. soft and puree
19. How often child eats sweets daily?	a. once a day      b. twice a day      c. three or more times a day      d. does not eat sweets
20. Does your child consume high – energy, natural and syntetic supplements with sugar?	a. yes      b. no

filled numbered 70 (42.4%) (Table 3). On average, every child had 5.8 affected deciduous teeth. In the structure of dmft, with a total of 70 teeth, 64 (91.4%) had caries, 3 (4.3%) teeth were extracted and 3 (4.3%) were sealed. Comparing dental caries status and the structure of dmft between EB and healthy control children, there were no statistically significant differences (Table 4).

From the twelve patients, eight (66.7%) had permanent dentition, and affected teeth, while four (33.3%) had all healthy teeth. Of a total of 225 permanent teeth, there were 171 (76%) healthy teeth and 54 (24%) listed with caries or as extractions or filled teeth (Table 3). Every child had an average of 5 affected permanent teeth. Of a total of 54 teeth in the structure of DMFT, there were 45 (83.3%) caries, 6 (11.1%) extractions and 3 (5.6%) sealed teeth (Table 4). There was a statistically significant difference in the distribution of caries, extracted and filled teeth between children with EB and the healthy control group. In children in both groups, the

most common symptom was dental caries but there were more extracted teeth in the EB group, and more filled teeth in the control group (Table 4).

#### Oral hygiene status

In primary dentition, the average value of the modified plaque index was significantly higher in children with EB in relation to the control group and was  $1.4 \pm 1.14$  (minimum was 0 and maximum was 3). The modified calculus index in children with EB on average had a value of  $0.7 \pm 1$  (minimum was 0 and the maximum was 2). None of the children in the control group had calculus on deciduous teeth. In EB children, on permanent teeth, the plaque index showed significantly higher values (1.3 to 2.6; an average of  $2 \pm 0.4$ ) as compared to the healthy children of the control group. The calculus index was also significantly higher in the study group (0.7 to 2.5; an average of  $1.6 \pm 0.6$ ) than in the control group (Table 5).

**Table 3**

Dentition	Teeth (%)				dmft/DMFT $\bar{x} \pm SD$
	Caries free	Decayed (caries)	Missing	Filled	
Primary					
affected subjects	57.6	38.8	1.82	1.82	$5.83 \pm 5.69$
control subjects	77.8	19.6	0.0	2.5	$3.50 \pm 3.21$
Permanent					
affected subjects	76	20	2.7	1.3	$4.50 \pm 4.56$
control subjects	86.8	9.2	0.4	3.5	$2.50 \pm 2.54$

**DMFT – the decayed, missing, filled teeth index; dmft – a variation of DMFT that is applied to the primary dentition;  $\bar{x}$  – mean value; SD – standard deviation.**

**Table 4**

Dentition	dmft/DMFT n	Teeth, n (%)		
		decayed	missing	filled
Primary				
affected	70	64 (91.4)	3 (4.3)	3 (4.3)
control	35	31 (88.6)	0(0)	4 (11.4)
<i>p</i>			0.189*	
Permanent				
affected	54	45 (83.3)	6 (11.1)	3 (5.6)
control	30	21 (70.0)	1 (3.3)	8 (26.7)
<i>p</i>			0.021*	

\* $\chi^2$ -test

For abbreviations see under Table 3.

**Table 5**

Characteristic	Group		<i>p</i>
	affected	control	
Modified plaque index <sup>1</sup> , $\bar{x} \pm SD$	$1.54 \pm 0.96$	$0.26 \pm 0.27$	0.011 <sup>§</sup>
Modified calculus index <sup>1</sup> , $\bar{x} \pm SD$	$0.78 \pm 0.98$		
Plaque index <sup>1</sup> , $\bar{x} \pm SD$	$1.97 \pm 0.44$	$0.26 \pm 0.25$	< 0.001 <sup>§</sup>
Calculus index <sup>1</sup> , $\bar{x} \pm SD$	$1.59 \pm 0.57$	$0.10 \pm 0.21$	< 0.001 <sup>§</sup>
Presence of gingivitis, n (%)	16 (94.1)	5 (29.4)	< 0.001*

\* $\chi^2$ -test; <sup>§</sup>Mann-Whitney U test.

<sup>1</sup>Note: – as described by Greene and Vermillion<sup>17</sup>.

For abbreviations see under Table 3.

Figure 1 depicts much dental caries, untreated teeth and hard and soft tissue deposits in a child patient with *epidermolysis bullosa*.



**Fig. 1 – State of hard and soft tissue in patient with *epidermolysis bullosa*.**

A child with pseudosyndactyly and reduced, manual dexterity that result in difficulties to maintain a daily oral hygiene routine is shown in Figure 2.



**Fig. 2 – Pseudosyndactyly in a patient with *epidermolysis bullosa*.**

#### *Oral soft tissues, gingiva and mouth opening capacity*

From the 17 patients suffering from EB, 13 (76.5%) had *microstomia* (Figure 3). The average mouth opening capacity was measured to be  $40.1 \pm 6.6$  mm with a maximal value at 49 mm and a minimal value at 24 mm. Fifteen (88.2%) patients had *ankyloglossia*. Vestibular obliteration was observed in 10 (58.8%) patients. *Bullae* were present in 10 (58.8%) patients and the absence of lingual *papillae* and palatal *rugae* were diagnosed in 14 (82.4%) of the children. Perioral *bullae* and scars were present in 12 (70.6%) patients.

Gum disease (*gingivitis catarrhalis*) was diagnosed in 16 (94.1%) of the patients with only the youngest patient (aged one year) marked as having healthy gums (5.9%). The-

re was a statistically significant difference in the frequency of gum disease among the group of children with EB and the control group of healthy children (Table 5).



**Fig. 3 – Microstomia in a patient with *epidermolysis bullosa*.**

#### *The level of knowledge and the opinions of parents about the quality of dental care of EB children*

Results obtained from analyzing the questionnaires showed that after the diagnosis of EB in children had been set, none of the parents had been recommended to seek advice from dentists on the prevention of oral diseases. Nine (52.9%) patients do not have a regular dentist and go to the dental clinic only when problems appear. The other eight (47.1%) patients had had preventive examinations and were given tips on how to maintain oral health. All parents expressed the need for a dentist who is trained to work with children with EB because almost half (47.1%) of the respondents were only able to find dentists who did not dare to carry out intervention due to insufficient knowledge and experience in this area. Only one (5.9%) patient had been administered the local application of highly concentrated fluoride.

Regarding the maintenance of oral hygiene, approximately half (52.9%) of respondents brush their teeth two times a day, five (29.4%) more than 2 times, and three (17.7%) once a day and also not every day. But more than half (64.7%) of the affected children do not brush their teeth at the appropriate times (in the morning before breakfast and in the evening before bedtime). Toothpaste with fluoride was being used by 65.7% of the children and toothpaste without fluoride was being used by 35.3% of the children. None of the patients were using methods for dental plaque identification in order to control and improve oral hygiene. Eleven (64.7%) patients were using soft brushes, four (23.5%) patients were using medium-hard brushes, and four (23.5%) were using special toothbrushes. Two (11.8%) patients were not aware that toothbrushes come in varying degrees of hardness.

When unable to brush their teeth after a meal, 12 (70.7%) of the patients rinse their mouth with water, and 5 (29.4%) do not. Two (11.8%) patients had been using oral rinses containing fluoride, and five (29.4%) had been using herbal rinses.

Ten (58.8%) children were found not to use oral rinses. Also, none of the patients use antiplaque rinses with chlorhexidine.

Concerning nutrition, all of the children have more than 5 meals *per* day which last an average of 20–30 minutes. Thirteen (76.5%) of the children consume soft and puree food and four (23.5%) children consume solid but not sharp food (chips, popcorn, crusts of bread, etc.), which might hurt fragile oral mucosa. Eight (46.1%) respondents eat sweets twice a day, and a further eight eat sweets three or more times *per* day. Sixteen of the (94.1%) children consume high-energy, natural and synthetic supplements with sugar. Fourteen patients were shown to be uninformed about medications and supplements that do not contain sugar.

## Discussion

This research has shown that children with EB have high risk prevalence for caries and periodontal disease, and that they have very poor oral hygiene. The children were shown to a statistically significant degree to present with more caries, with untreated teeth and significantly more soft and hard deposits on teeth compared to the children in the control group.

Although the statistically significant differences between the groups in this study were not found in the distribution of cavities that objectively existed, the significance was not expressed because of the small number of patients. It was observed (Table 1) that children with EB have double the percent of affected teeth as compared to healthy children. It is significant that in the category of teeth with caries, two thirds of the caries presented in the EB children were graded as the severe forms of dental caries (deep caries and complications) which was not the case in the control group of healthy children. Anomalies of tooth structure in terms of enamel hypoplasia were also recorded in two (11.8%) of the EB children.

A small percentage of filled teeth can be explained by the neglect of oral health which might be caused by other symptoms of these diseases. Another reason might be the insufficient experience and knowledge of most dentists in how to treat children with EB and the objective difficulties involved with treatment (working with a narrowed oral aperture and therefore a narrowed field of vision). The children cannot hold their mouth open easily and cotton rolls placed with difficulty because of the vestibular obliteration. For the same reason, it is often impossible to place the dental mirror correctly to separate soft tissues from a tooth, and dental work can also cause the appearance of bullae, etc.

The main reasons for poor oral hygiene include but are not limited to: reduced mobility of the tongue caused by ankyloglossia, difficulty in chewing and swallowing with prolonged retention of food in the mouth, consumption of puree and sticky food, etc.<sup>6, 15</sup>. This is supported by the fact that sixteen of the seventeen patients had inflamed gingiva, and the percentage of caries was high in primary and in permanent dentition. A daily oral hygiene routine is difficult to maintain according to some authors because of the reduced manual dexterity (scarring and pseudosyndactyly), the presence of *bullae*, microstomia, difficulty in opening the mouth, ankyloglossia, shallow or obliterated vestibuli and ena-

mel hypoplasia<sup>6, 15, 18</sup>, which was confirmed in this study. When access to some teeth is difficult the use of single brushes with a small head and long handle is usually recommended. Use of an ultrasonic toothbrush and dental water jet (waterpik) could be greatly beneficial, but despite recommendations, patients expressed difficulty with obtaining such devices due to economic concerns.

If the use of a toothbrush is not possible for any reason, the advice given to parents is to wrap wet gauze around the finger and to gently clean the accessible surfaces of the teeth. That parents instill a regular oral hygiene routine in children is vital<sup>12, 19</sup>. Many parents are afraid, however, that during brushing they could cause blisters, and therefore do not regularly apply oral hygiene to their children.

An analysis of nutritional habits revealed that the EB patients spent about 150 minutes *per* day eating. Most of them consume puree and soft food and eat sweets at least twice daily. Clearance of food is prolonged which, combined with frequent meals rich in calories, leads to the rapid development of caries and early tooth decay (loss)<sup>12</sup>. It is necessary for EB children to have a diet with an increased protein intake by 15–100%, and an overall calorie intake increase of 20–50%. As a result, such patients have frequent high-calorie and high-protein meals<sup>20</sup>.

Questionnaires collected from the parents indicated that children suffering from EB drink various natural and artificial drinks very slowly (gulp by gulp) and that it takes them almost 20 minutes. Additionally, snacks tend to be the sticky foods which are slowly eliminated from the mouth. It is hard to influence nutrition habits because of the specific nutritional needs of these patients<sup>21</sup>. However, some small changes can be made in the vein of reducing the frequency of taking food between main meals, consuming less cariogenic food and sugar-free, high-protein energy substitutes and eating sweets once a day after a meal. Most patients in this study stated that after eating, when they are not able to brush their teeth, they rinse their mouth with water. However, the effectiveness of rinsing is significantly reduced due to the scarring changes of oral soft tissues and the reduced mobility of mucosa and muscle hypotonia. These symptoms taken together cause this measure to have almost no effect.

Analysis of preventive measures carried out on children with EB showed that no child had sealed fissures prior to participation in this study and only one patient had had an application of a high dose varnish of fluorides. These prophylactic measures, as well as the identification of dental plaque and the mechanical removal of soft deposits and teeth calculus with ultrasonic cleaners, were subsequently done at the Clinic for Pediatric and Preventive Dentistry after a certain level of cooperation with children was achieved. The advice was given regarding oral hygiene, correcting bad habits in nutrition and using products with fluoride as well as implementing oral rinses that prevent accumulation of dental plaque and tartar.

For children who are able to spit, it is advised that toothpaste with higher fluoride concentrations should be applied regularly<sup>22</sup>. Given the high percentage of caries and gingivitis, the recommendations received at the clinic included the

daily use of a solution of chlorhexidine (0.05%) and fluoride (223 ppmF). Excellent results have shown to be achieved using higher concentrations of chlorhexidine for a period of one week every 2–3 months<sup>19</sup>. It should be noted that prior to this study none of the respondents did not use chlorhexidine, which is one of the most effective agents for chemical control of dental plaque. When a child does not know how to spit out, parents were advised to apply rinse to a gauze or cotton swab and coat the teeth and gums. Children with EB should use solutions without alcohol and menthol as rinses can irritate the oral mucosa<sup>21</sup>. Dental visits were scheduled for every 3 months. Although all patients received information about basic and additional measures to maintain and improve oral health, few have succeeded to implement them in a daily routine. Because of that, it is necessary to have constant monitoring and motivation of child and parent; otherwise, there is an inevitable neglect of oral health care.

This study showed that dental therapy interventions can be conducted in children with EB. On their first visits to the clinic, the parents wanted restorations to be done rather than preventive measures to be performed on their children. The reason for that is that they did not come on time to regular check-ups in the past and many of the children already had severe dental problems. Dental treatment was difficult because of microstomia, shallow vestibuli and the appearance of *bullae* at the slightest application of pressure on the oral mucosa. *Bullae* were perforated with thin sterile needles of syringes, to avoid the formation of scar tissue. With cautious work "in the air" and with frequent interruptions as well as the application of local anesthesia almost all conservative

treatments were made and tooth extractions completed. It should be noted that the indications for endodontic treatment in children with EB are limited to the front teeth (due to microstomia) and that for the more complicated tooth extraction it is necessary to use general anesthesia which requires special conditions and additional trained staff.

### Conclusion

Children with EB account for a group of high-risk patients for developing caries and periodontal disease. Of all patients with a medical risk, the toughest dental work is with this category of patients due to the sensitivity of the oral mucosa, so only a small number of dentists is available to these patients even in the performance of simpler preventive dental procedures. The parents of children are not sufficiently informed about the importance of the early application of preventive measures to preserve oral health and they find that their children do not have adequate dental care. In order to alleviate this problem there must be a proper cooperation protocol between the treating physician and the dentist. After an EB diagnosis is made, the child and parents need to be in contact with a qualified dentist who will educate them and take preventive measures to preserve oral health as light and non-aggressive as possible: the treatment of choice for all patients at risk, especially for children with EB. It is necessary even within the specialization of pediatric and preventive dentistry to organize subspecialty training for dentists who are to work with patients affected with EB.

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