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Dental Status and Periodontal Health of Patients with Phenylketonuria in Latvia

Dentalni status i parodontološko zdravlje pacijenata s fenilketonurijom u Latviji

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Abstract

Objectives: Phenylketonuria (PKU) is an autosomal recessive inherited disorder of phenylalanine metabolism resulting from a deficiency of phenylalanine hydroxylase. The aim of this study was to evaluate the dental status and periodontal health of PKU patients in Latvia. **Material and Methods:** Forty-five PKU patients and age/sex-matched controls were recruited for this cross-sectional study. Their anamnestic data, periodontal health and dental status were assessed by one experienced dentist. **Results:** Dental and periodontal clinical evaluation revealed that the median number of filled teeth was significantly smaller among PKU patients compared to the control group ($p=0.021$). PKU patients had a significantly larger median number of carious teeth than their healthy counterparts ($p<0.001$). Significant differences between the PKU and control groups were observed for several oral hygiene indices ($p<0.001$): Silness-Löe plaque index, OR=29.3 (95% CI: 3.7–232.4); CPITN index, OR=35.2 (95% CI: 4.5–278.3); Greene-Vermillion index, OR=10.2 (95% CI: 2.8–38.0); calculus removal necessity, OR=12.3 (95% CI: 3.3–45.4). **Conclusion:** Dental status and periodontal health of PKU patients was found to be significantly inferior compared to healthy controls. This is likely due to the regular consumption of PKU formula and the difficulties which mentally and/or physically disabled PKU patients experience with their oral hygiene. To prevent tooth decay and periodontal disease, PKU patients should visit a professional oral hygienist every three to six months. Furthermore, they should adopt the habit of rinsing their mouth with water immediately after consuming PKU formula to counteract the acidity in their oral cavity.

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Introduction

Phenylketonuria (PKU) is an autosomal recessive inherited disorder of phenylalanine (Phe) metabolism resulting from a deficiency of phenylalanine hydroxylase (prevalence 1:10,000), (1,2). Untreated, PKU can lead to intellectual disability, seizures, behavioural problems and mental disorders (3). The treatment of PKU entails following a strict low-protein diet and consumption of Phe-free amino acid formula. For all PKU patients, medical nutrition therapy is the primary form of treatment as it is the only way to avoid ingestion of the amino acid Phe while still meeting their protein intake needs (4). However, although this formula is essential for the wellbeing of PKU patients, it can negatively impact their oral health as it contains a high amount of complex carbohy-

Uvod

Fenilketonurija (PKU) je autosomno recesivni nasljedni poremećaj metabolizma fenilalanina (Phe), a rezultat je nedostatka fenilalanin hidroksilaze (prevalencija 1:10 000), (1, 2). Neliječen, PKU može rezultirati intelektualnim invaliditetom, napadajima i mentalnim poremećajima te poremećajima u ponašanju (3). Liječenje podrazumijeva strogu dijetu s malo proteina i konzumaciju formule bez Phe aminokiselina. Za sve pacijente s PKU-om je terapija medicinskom prehranom primarni oblik liječenja jer je to jedini način da se izbjegne unos aminokiseline Phe, a da su pritom zadovoljene njihove potrebe za unosom proteina (4). No iako je ta formula prijeko potrebna za dobrobit pacijenata s PKU-om, može negativno utjecati na oralno zdravlje jer sadržava veliku količinu

drates. There are two types of complex carbohydrates – starch and fibre. Salivary and bacterial amylases in human saliva hydrolyse starches into maltose, maltotriose and low-molecular dextrans (5). In high concentrations, these by-products serve as excellent substrates for bacteria that are used in acid production. An increased acidity in oral cavity too often or for prolonged periods of time results in demineralisation of tooth structures. This process is known as dental caries, and all PKU patients are susceptible to higher levels of caries activity due to their dietary needs (5).

Furthermore, patients with mental and/or developmental problems, as seen in PKU patients diagnosed later in life or in cases of poor diet compliance, are likely to find it difficult to sustain adequate dental hygiene (6). Thus, based on their nutritional needs and possible inability to properly brush their teeth, the oral health of PKU patients is likely to be inferior to healthy controls.

Oral health encompasses a range of diseases and conditions. The current study focused on assessment of subjects' dental status (identification of decayed, missing and filled teeth), oral hygiene habits (frequency of tooth brushing, use of additional oral hygiene tools and fluoride supplements) and evaluation of subjects' periodontal health by means of clinical examination and oral hygiene indices. Periodontal disease is a multifactorial chronic inflammatory condition of periodontium (the supporting structures of teeth) caused mainly by dysbiosis of oral microflora, in which periodontal pathogens thrive. If left untreated, periodontal disease results in irreversible localised or generalised alveolar bone loss due to constant inflammatory processes in periodontium. Bacteria are the main etiological factor of periodontal disease; however, the severity of periodontal lesions can be influenced by numerous environmental factors and acquired diseases. There can also be a certain degree of genetic predisposition to periodontal disease (7,8). In most cases, incorrect and/or irregular tooth brushing coupled with a failure to use interdental cleaning products is the direct cause of periodontal disease development since perfect conditions for bacteria to flourish in persistent dental plaque and calculus are thus created (9,10). Another important factor that influences oral health is salivary flow. Saliva is a very important protective factor in oral cavity. Individuals with a low unstimulated salivary flow rate are more susceptible to dental caries because their saliva contains less of the main salivary buffer bicarbonate, as its concentration in saliva is proportional to flow rate. Lowered salivary production also predisposes individuals to xerostomia (dry mouth syndrome) and ensures that clearance of food from the mouth is delayed (11).

Since there is only a limited number of reports on the oral health of PKU patients, one of the main objectives of the present study was to investigate the dental status and periodontal health of PKU patients aged 12 years or older in Latvia. An Evaluation of the dental-medical history and the assessment of their permanent teeth and periodontal tissues were carried out.

složenih ugljikohidrata. Dvije su vrste složenih ugljikohidrata – škrob i vlakna. Slina i bakterijske amilaze hidroliziraju škrob u maltozu, maltotriozu i niskomolekularne dekstrine (5). Ti nusproizvodi u visokim koncentracijama izvrsni su supstrati za bakterije u proizvodnji kiselina. Prečesto ili dulje povećana kiselost u usnoj šupljini demineralizira zubne strukture. Taj je proces poznat kao zubni karijes, a svi pacijenti s PKU-om, zbog svojih prehranbenih potreba, predisponirani su za višu razinu karijesne aktivnosti (5).

Nadalje, pacijentima s mentalnim i/ili razvojnim problemima – poput pacijenata s PKU-om dijagnosticiranim kasnije u životu ili u slučaju lošega pridržavanja prehrane – vjerojatno će biti teško održavati odgovarajuću higijenu zuba (6). Zato će, na temelju njihovih prehranbenih potreba i nemogućnosti pravilnoga pranja zuba, oralno zdravlje tih pacijenata vjerojatno biti lošije u usporedbi sa zdravom kontrolnom skupinom.

Oralno zdravlje obuhvaća niz bolesti i stanja. Aktualno istraživanje bilo je usmjereno na procjenu dentalnoga statusa ispitanika (prepoznavanje zuba s karijesom, izvađenih zuba i zuba s ispunima), navike oralne higijene (učestalost pranja zuba, korištenje dodatnih oralnih higijenskih sredstava i dodataka fluora) i procjenu parodontološkoga zdravlja ispitanika s pomoću kliničkog pregleda i indeksa oralne higijene. Parodontitis je multifaktorijalno kronično upalno stanje parodonta (potpornih struktura zuba) prouzročeno uglavnom disbiozom oralne mikroflore u kojoj se razvijaju parodontni patogeni. Ako se ne liječi, parodontitis rezultira nepovratnim lokaliziranim ili generaliziranim gubitkom alveolarne kosti zbog stalnih upalnih procesa u parodontu. Bakterije su glavni etiološki čimbenik parodontitisa, no na težinu parodontoloških lezija mogu utjecati mnogobrojni okolišni čimbenici i stečene bolesti. Također može postojati određeni stupanj genetske predispozicije (7, 8). U većini slučajeva, nepravilno i/ili neredovito pranje zuba u kombinaciji s nekorištenjem interdentalnih pomagala za čišćenje, izravni je uzrok pojave parodontitisa jer se stvaraju savršeni uvjeti za razvoj bakterija u zubnome plaku i kamencu (9, 10). Drugi važan čimbenik koji utječe na oralno zdravlje jest protok sline. Naime, slina je vrlo važan zaštitni čimbenik u usnoj šupljini. Osobe s niskim nestimuliranim protokom sline osjetljivije su na zubni karijes jer je u njihovoj slini manje puferskoga bikarbonata s obzirom na to da je njegova koncentracija proporcionalna brzini protoka. Smanjena proizvodnja sline također predisponira pojedince na kserostomiju (sindrom suhih usta) i pogoduje odgođenom uklanjanju hrane iz usta (11).

Budući da postoji samo ograničeni broj izvješća o oralnome zdravlju pacijenata s PKU-om, jedan od glavnih ciljeva ovoga rada bio je u Latviji istražiti dentalni status i parodontološko zdravlje takvih pacijenata u dobi od 12 ili više godina. Obavljena je procjena medicinske i stomatološke anamneze pacijenata te procjena njihovih trajnih zuba i parodonta.

Material and methods

Ethics

Approval for this study was granted by the Central Medical Ethics Committee and Genome Research Council of Latvia prior to data collection. The study was conducted according to the Helsinki Declaration. All possible risks, objectives, and benefits of involvement in the study were carefully explained to every participant or the parents of minor age participants and mentally disabled patients. Before participation in research, each participant or their representative(s) filled out a questionnaire (Appendix 1).

Materijali i metode

Etičko odobrenje

Odobrenje za istraživanje dali su Središnji odbor za medicinsku etiku i Vijeće za istraživanje genoma Latvije prije prikupljanja podataka. Istraživanje je provedeno prema Helsinškoj deklaraciji. Svi mogući rizici, ciljevi i koristi od uključivanja u istraživanje pomno su objašnjeni svakom sudioniku ili roditeljima maloljetnih sudionika i mentalno invalidnih pacijenata. Svaki sudionik ili njegov predstavnik na početku je ispunio upitnik (prilog 1.).

Appendix 1 Dodatak 1.

The questionnaire contained inquiries about: • Upitnik je sadržavao pitanja o:

1. General health medical history. • općoj zdravstvenoj anamnezi
2. Dental hygiene habits (frequency of tooth brushing, flossing, use of mouthwash and fluoride supplements). • oralno-higijenskim navikama (učestalost pranja zuba, korištenje konca, korištenje otopina za ispiranje usta i dodatak fluora)
3. Frequency of visits to the dentist and dental hygienist. • učestalosti posjeta stomatologu i dentalnom higijeničaru
4. Overall satisfaction with their oral health. • sveukupnom zadovoljstvu oralnim zdravljem
5. Daily water intake. • dnevnom unosu vode
6. Nutritional habits (frequency of meals, preferred foods at main mealtimes, snacking habits). • prehranbenim navikama (učestalost obroka, preferirana hrana u vrijeme glavnih obroka, navike grickanja).

PKU patients additionally were asked about: • Pacijentima s PKU-om dodatno su postavljena pitanja o:

7. Their PKU diagnosis (at what age the diagnosis was established). • njihovoj dijagnozi PKU-a (u kojoj dobi je dijagnoza postavljena).
8. Their adherence to PKU diet (intake of Phe-free medical formula and other low-Phe foods, as well as how successfully patients are avoiding high-Phe foods). • pridržavanju PKU prehrane (unos medicinske formule bez Phe i druge hrane s niskim sadržajem Phe, te koliko uspješno izbjegavaju hranu s visokim sadržajem Phe).
9. Their current and usual plasma Phe concentrations. • njihovoj trenutačnoj i uobičajenoj koncentraciji Phe u plazmi.

Study design

Forty-five PKU patients (62.2% females and 37.8% males) of 50 PKU diagnosed in Latvia between the ages 12 and 53 years (median=22) who agreed to participate in this cross-sectional study were recruited. 35 PKU patients were diagnosed with PKU immediately after birth, and 10 participants had delayed diagnoses. The healthy control group consisted of 45 age- and sex-matched individuals.

Salivary sample collection

Upon arrival to the dental office all participants were instructed to have breakfast and brush their teeth in the morning as usual but not later than 2 hours before sample collection time. The questionnaire (Appendix 1) was followed by salivary sample collection to determine basal salivary secretion rate. Participants were provided a graduated plastic tube for sample collection and a quiet, private space. Saliva sample was taken by unstimulated drain method which is easy and safe. No specific procedures were required other than sitting with the head slightly down and spitting saliva spontaneously secreted into the tube. Participants were timed and instructed to keep spitting until 10 ml mark was reached on the tube. Afterwards, the 10 ml were divided by number of minutes required for the participant to provide the sample and to determine whether their salivary flow is within the normal range of 0.3 – 0.4 ml/min. In Table 1, this is displayed as > 60 seconds (it took participants more than 60 seconds to secrete 0.3 – 0.4 ml of saliva, which indicates decreased salivary flow)

Studijski dizajn

Sudjelovalo je 45 pacijenata s PKU-om (62,2 % žena i 37,8 % muškaraca) od njih 50 dijagnosticiranih u Latviji u dobi od 12 do 53 godine (medijan = 22) koji su pristali sudjelovati u presječnom istraživanju. Kod njih 35 pacijenata bolest je dijagnosticirana odmah nakon rođenja, a 10 je imalo odgođenu dijagnozu. Zdravu kontrolnu skupinu činilo je 45 osoba podudarne dobi i spola.

Prikupljanje uzoraka sline

Poslije dolaska u stomatološku ordinaciju svim je sudionicima rečeno da doručkuju i ujutro operu zube na uobičajeni način, ali najkasnije dva sata prije uzimanja uzorka. Nakon ispunjavanja upitnika (dodatak 1.) slijedilo je uzimanje uzoraka sline da bi se odredila njezina bazalna brzina izlučivanja. Svi su dobili graduiranu plastičnu epruvetu za prikupljanje uzoraka i miran, privatni prostor. Uzorak sline uzet je nestimuliranom metodom, jednostavno i sigurno, za što nisu potrebni posebni zahvati osim sjedenja pognute glave i pljuvanja sline koja se spontano izlučuje u epruvetu. Sudionicima je određeno vrijeme i naloženo da nastave pljuvati dok se na epruveti ne postigne oznaka od 10 mL. Nakon toga je tih 10 mL podijeljeno s brojem minuta koje su sudioniku bile potrebne da prikupi uzorak i utvrdi je li njihov protok sline unutar normalnoga raspona od 0,3 do 0,4 mL/min. (U tablici 1. to je prikazano kao > 60 sekunda – sudionicima je trebalo više od 60 sekunda da izluče 0,3 – 0,4 mL sline, što upućuje na smanjeni protok sline) i < 60 sekunda (sudionicima

and < 60 seconds (it took participants less than 60 seconds to secrete 0.3 – 0.4 ml of saliva, which indicates salivary flow within normal range).

Clinical examination of teeth and periodontal tissues

Salivary sample collection was followed by clinical examination of teeth and periodontal tissues. Clinical examination was carried out by a single dentist under appropriate and uniform lighting conditions. Dental status was assessed by identifying decayed teeth, missing teeth and filled surfaces of teeth (DMFS index) using visuo-tactile dental examination with a sharp dental probe, mirror, 3-1 syringe and dental magnifying loupes worn by the dentist. Several indices were used to assess oral hygiene and gingival health: CPITN index, Silness-Löe plaque index and Greene-Vermilion index. Values for these indices were determined by assessing the presence and abundance of plaque and calculus for specific teeth, measuring the depth of gingival sulci and/or periodontal pockets and evaluating gingival bleeding on probing with a periodontal probe.

Patients with CPITN scores of 1 and 2 were labelled as gingivitis patients, while patients with CPITN scores of 3 and 4 were classified as possible risk of periodontal disease.

Statistical analysis

A statistical analysis was conducted using SPSS for Windows. A risk analysis was performed, and the magnitude of the risk was estimated by the odds ratio (OR) and its 95% confidence interval (95% CI). A p-value of <0.05 was considered to be an indicator of a statistically significant result.

Results

In total, 45 PKU patients and 45 healthy controls were examined. Table 1 represents the oral hygiene habits, professional oral hygiene necessity and basal salivary secretion rate for both PKU and control groups. The most striking difference between the two groups was observed in the frequency of tooth brushing; almost all the healthy controls (95.6%) reported brushing their teeth twice a day, whereas there were only 60% of PKU patients ($p < 0.001$). There were also marked differences between the two groups in the use of dental floss and mouthwash. A smaller number of PKU patients reported using dental floss compared with control individuals (15.6% vs. 60.0%); however, a larger number of PKU patients used mouthwash (8.9% vs. 2.2%). Notably, a significantly greater number of PKU patients reported not using any interdental cleaning products (75.6% vs. 37.8%; $p < 0.001$). A significant difference between the two groups was also observed in professional oral hygiene necessity, with more PKU patients requiring this procedure (93.3% vs. 53.3%) (OR=12.3, 95% CI: 3.3–45.4; $p < 0.001$). The control group, on average, had a higher basal salivary secretion rate (63.6% of control group patients had a salivary flow rate within normal range, compared to 40% of PKU patients, $p = 0.03$).

Clinical examination of teeth revealed a significantly smaller median number of filled teeth among PKU patients compared with control individuals ($p < 0.05$) (Table 2). In contrast, PKU patients had a significantly larger medi-

je trebalo manje od 60 sekunda da izluče 0,3 – 0,4 mL sline, što upućuje na protok sline unutar normalnoga raspona).

Klinički pregled zuba i parodonta

Nakon uzimanja uzoraka sline slijedio je klinički pregled zuba i parodonta. Obavio ga je jedan kliničar u odgovarajućim i ujednačenim uvjetima osvjetljenja. Dentalni status ocjenjivan je identifikacijom zuba s karijesom, zuba koji nedostaju i ploha s ispunima (DMFS indeks) vizualno-taktilnim stomatološkim pregledom oštrom dentalnom sondom, ogledalom, 3 – 1 štrcaljkom i dentalnim povećalom. Za procjenu oralne higijene i zdravlja gingive upotrijebljeno je nekoliko indeksa: CPITN indeks, Silness-Löeov indeks plaka i Greene-Vermillionov indeks. Vrijednosti tih indeksa određene su procjenom prisutnosti i količine plaka i kamenca za određene zube, mjerenjem dubine gingivnih brazda i/ili parodontnih džepova i procjenom krvarenja gingive pri sondiranju parodontološkom sondom.

Pacijenti s CPITN ocjenama 1 i 2 označeni su kao pacijenti s gingivitisom, a oni s CPITN ocjenama 3 i 4 klasificirani kao pacijenti s mogućim rizikom od parodontitisa.

Statistička analiza

Statistička analiza obavljena u programu SPSS-a za Windows. Provedena je analiza rizika, a veličina rizika procijenjena je omjerom vjerojatnosti (OR) i njegovim 95-postotnim intervalom pouzdanosti (95 % CI). P-vrijednost < 0,05 smatrana je pokazateljem statistički značajnoga rezultata.

Rezultati

Ukupno je pregledano 45 pacijenata s PKU-om i 45 zdravih kontrola. Tablica 1. prikazuje navike oralne higijene, potrebu za profesionalnom oralnom higijenom i bazalnu sekreciju sline za pacijente s PKU-om i kontrolnu skupinu. Najdovoljivija razlika između dviju skupina uočena je u učestalosti pranja zuba – gotovo svi zdravi ispitanici iz kontrolne skupine (95,6 %) rekli su da peru zube dva puta na dan, dok je to izjavilo samo 60 % pacijenata s PKU-om ($p < 0,001$). Također su postojale značajne razlike između dviju skupina u korištenju zubnoga konca i vode za ispiranje usta. Manje pacijenata s PKU-om koristilo se zubnim koncem u usporedbi s kontrolnom skupinom (15,6 % prema 60,0 %), ali više pacijenata s PKU-om upotrebljavalo je tekućinu za ispiranje usta (8,9 % prema 2,2 %). Zanimljivo je da je znatno više pacijenata s PKU-om izjavilo da ne upotrebljavaju nikakve proizvode za interdentalno čišćenje (75,6 % prema 37,8 %; $p < 0,001$). Značajna razlika između dviju skupina zabilježena je također u potrebi za profesionalnom oralnom higijenom, s tim da je više pacijenata s PKU-om zahtijevalo taj postupak (93,3 % prema 53,3 %) (OR = 12,3, 95 % CI: 3,3 – 45,4; $p < 0,001$). Kontrolna skupina imala je u prosjeku višu stopu bazalnoga izlučivanja sline (63,6 % ispitanika iz kontrolne skupine imalo je brzinu protoka sline unutar normalnoga raspona, u usporedbi s 40 % ispitanika s PKU-om, $p = 0,03$).

Kliničkim pregledom zuba ustanovljen je značajno manji medijan broja zuba s ispunima kod pacijenata s PKU-om

Table 1 Oral hygiene habits, professional oral hygiene necessity and basal salivary secretion rate in PKU patients and control group.**Tablica 1.** Oralno-higijenske navike, potreba za profesionalnom oralnom higijenom i bazalno izlučivanje sline kod pacijenata s PKU-om i u kontrolnoj skupini

		Controls • Kontrolna skupina (n=45)	PKU cases • Pacijenti s PKU-om (n=45)	Total • Ukupno (n=90)	OR (95% CI)	p-value* • p-vrijednost*
Frequency of tooth brushing • Učestalost pranja zuba	Do not brush • Ne pere	0 0%	5 11.1%	5 5.6%	14.3 (3.1-66.7)	<0.001
	Once per day • Jedanput na dan	2 4.4%	13 28.9%	15 16.7%		
	Twice per day • Dvaput na dan	43 95.6%	27 60.0%	70 77.8%	ref	
Interdental cleaning products • Proizvodi za interdentalno čišćenje	Do not use • Ne upotrebljava	17 37.8%	34 75.6%	51 56.7%	5.1 (2.05-12.6)	<0.001
	Dental floss • Zubni konac	27 60.0%	7 15.6%	34 37.8%	ref	
	Mouthwash • Otopina za ispiranje	1 2.2%	4 8.9%	5 5.6%		
Professional oral hygiene necessity • Potreba za profesionalnom oralnom higijenom	Not necessary • Nije potrebno	21 46.7%	3 6.7%	24 26.7%	12.3 (3.3-45.4)	
	Necessary • Potrebno	24 53.3%	42 93.3%	66 73.3%		
Basal salivary secretion rate • Bazalna brzina izlučivanja sline	< 60 seconds • < 60 sekunda	28 63.6%	18 40.0%	46 51.7%	2.6 (1.1-6.2)	0.03
	> 60 seconds • > 60 sekunda	16 36.4%	27 60.0%	43 48.3%		

*Fisher's exact test • Fisherov egzaktni test

Table 2 Number of filled, extracted and carious teeth in PKU patients and control group.**Tablica 2.** Broj zuba s ispunima, izvađenih zuba ili s karijesom kod pacijenata s PKU-om i u kontrolnoj skupini

	Controls • Kontrolna skupina (n=45) Median number (IQR) • Medijan (IQR)	PKU cases • Pacijenti s PKU-om (n=45) Median number (IQR) • Medijan (IQR)	p-value* • p-vrijednost*
Filled teeth • Zubi s ispunima	6 (6)	4 (8)	0.021
Extracted teeth • Izvađeni zubi	0 (1)	0 (2)	0.07
Carious teeth • Zubi s karijesom	1 (2)	4 (6)	<0.001

*Mann-Whitney U test • Mann-Whitneyev U test; IQR – interquartile range • interkvartilni raspon

an number of carious teeth than their control counterparts ($p < 0.001$) (Table 2).

The values of oral hygiene indices were significantly higher in PKU patients than in control individuals ($p < 0.001$; Appendices 2, 4, 5); Silness-Löe plaque index, OR=29.3 (95% CI: 3.7–232.4); CPITN index, OR=35.2 (95% CI: 4.5–278.3); Greene-Vermillion index, OR=10.2 (95% CI: 2.8–38.0).

Regarding oral health and time of PKU diagnosis, it can be recognized that delayed PKU diagnosis can increase the risk of periodontal disease development. 80% of patients with a delayed diagnosis were determined to already have or to be at risk of developing periodontal disease, compared to only 31.4% of PKU patients diagnosed before the age of two months (OR=8.7, 95% CI: 1.6–48.1; $p < 0.05$) (Appendix 3).

u usporedbi s kontrolnom skupinom ($p < 0,05$) (tablica 2.). Suprotno tomu, pacijenti s PKU-om imali su značajno veći medijan broja zuba s karijesom od svojih kontrolnih parova ($p < 0,001$) (tablica 2.).

Vrijednosti indeksa oralne higijene bile su značajno veće kod pacijenata s PKU-om nego u kontrolnoj skupini ($p < 0,001$; prilozi 2, 4, 5); Silness-Löeov indeks plaka, OR = 29,3 (95 % CI: 3,7 – 232,4); CPITN indeks, OR = 35,2 (95 % CI: 4,5 – 278,3); Greene-Vermillionov indeks, OR = 10,2 (95 % CI: 2,8 – 38,0).

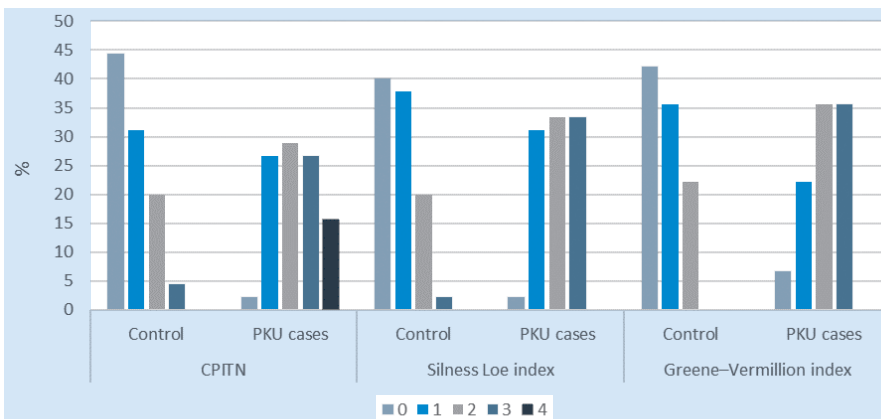
Kad je riječ o oralnome zdravlju i vremenu postavljanja dijagnoze PKU-a, uočava se da odgođena dijagnoza može povećati rizik od pojave parodontitisa. Ustanovljeno je da 80 % pacijenata s odgođenom dijagnozom već ima parodontitis ili je u opasnosti da će se pojaviti, u usporedbi sa samo 31,4 % pacijenata s PKU-om dijagnosticiranim prije navršena dva mjeseca (OR = 8,7, 95 % CI: 1,6 – 48,1; $p < 0,05$) (dodatak 3.).

Appendix 2 CPITN index of the PKU and healthy control group.**Dodatak 2.** CPITN indeks kod pacijenata s PKU-om i u zdravoj kontrolnoj skupini

	Control group • Kontrolna skupina (n=45)	PKU patients • Pacijenti s PKU-om (n=45)
0 (No present disease • nema bolesti)	20 44.4%	1 2.2%
1 (Gingival bleeding on probing • krvarenje gingive nakon sondiranja)	14 31.1%	12 26.7%
2 (Supragingival and/or subgingival calculus • supragingivni i/ili subgingivni kamenac)	9 20.0%	13 28.9%
3 (Pathological pocket depth 4-5 mm • patološka dubina džepova 4 – 5 mm)	2 4.4%	12 26.7%
4 (Pathological pocket depth >6mm • patološka dubina džepova > 6mm)	0 0.0%	7 15.6%
Total • Ukupno	45 100.0%	45 100.0%

Appendix 3 Diagnosis time of PKU patients in association with risk of periodontal disease.**Dodatak 3.** Povezanost vremena dijagnoze PKU-a i rizika od parodontitisa

		Diagnosis • Dijagnoza		Total • Ukupno
		Timely • Na vrijeme	Delayed • Odgodena	
Risk of Periodontal disease • Rizik od parodontitisa	No • Ne	24 68.6%	2 20.0%	26 57,8%
	Yes • Da	11 31.4%	8 80.0%	19 42,2%
Total • Ukupno		35 100,0%	10 100.0%	45 100.0%

**Appendix 4** CPITN, Silness & Löe, Greene-Vermillion index of the PKU and healthy control group.**Dodatak 4.** Indeksi CPITN, Silness-Löev, Greene-Vermillionov kod pacijenata s PKU-om i u zdravoj kontrolnoj skupini**Appendix 5** Silness & Löe, Greene- Vermillion index of PKU and healthy control group.**Dodatak 5.** Indeksi CPITN, Silness-Löev i Greene-Vermillionov kod pacijenata s PKU-om i u zdravoj kontrolnoj skupini

Characteristic	Control, N=45 ¹	PKU, N=45 ¹	p-value ²
Silness Löe index			<0.001
0	18 (40%)	1 (2.2%)	
1	17 (38%)	14 (31%)	
2	9 (20%)	15 (33%)	
3	1 (2.2%)	15 (33%)	
Greene-Vermillion index			<0.001
0	19 (42%)	3 (6.7%)	
1	16 (36%)	10 (22%)	
2	10 (22%)	16 (36%)	
3	0 (0%)	16 (36%)	

¹n(%) ²Pearson's Chi-squared test

Discussion

Prior to conducting this study, it was evident to the authors that the subject of oral health in PKU patients requires more in-depth research as very few articles on this subject are available at present, and the past articles are predominantly focused on the oral health of children. Furthermore, the available studies have reported contradictory results. For example, Kilpatrick and colleagues (12), examined 40 children with PKU and found no difference in the amount of dental caries compared with age/sex-matched healthy controls. However, significantly greater number of children with PKU exhibited the signs of tooth wear compared to their healthy counterparts (33% vs. 24%). More recently, in line with our results, Ballikaya et al. (13), reported higher caries prevalence among PKU patients. Nevertheless, it should be noted that their study primarily examined very young patients (1 to 5 years old) and only a small percentage (16.2%) of children was older than 11 years. Again, in accordance with our results, da Costa Silveira et al. (14), found that 75% of PKU patients were at high risk of caries development. They concluded that caries prevention and treatment were of major importance in dental care of these patients. One of the most extensive studies on this subject was conducted by Singh-Hüsgen and colleagues (15). They examined the oral health of 283 children with PKU and found that they experienced higher caries levels than healthy controls, which is a finding similar to the one presented in the current study for adults with PKU.

Our study showed that PKU patients have a decreased salivary secretion rate compared to the control group. Shimazaki et al. (16), examined the association of salivary flow rate with dental caries prevalence and periodontal status among 2,110 Japanese adults and suggested that individuals with lower salivary flow rates have higher risks for both dental caries and periodontal disease. Saliva is an important factor in a plethora of oral functions, such as mastication, swallowing, antimicrobial activity and cleaning action. Saliva also influences oral health both through its non-specific physio-chemical properties, as well as through more specific effects (17).

Significant finding in our study was an increased risk of periodontal disease in PKU patients. This is in line with other studies. For example, Lucas et al. (18), found a significantly greater mean plaque score for 41 PKU patients compared with controls. Ballikaya et al. (13), found moderate plaque accumulation and gingival inflammation in PKU patients, with nearly all of them requiring professional oral hygiene procedures due to excessive plaque and calculus. The scores for several oral hygiene indices were noticeably higher for our PKU patients compared to their controls, thus indicating an increased periodontal disease risk. However, it should be noted that the CPITN index has some limitations as it is based on gradual scoring. It lacks measurement of tooth mobility and attachment loss, which are important clinical symptoms of periodontal disease. The CPITN index should not be the sole approach for diagnosing periodontal disease. It is merely a tool often used in epidemiological studies to determine if a patient is at risk of periodontal problems (19). The main

Rasprava

Prije provedbe ovog istraživanja, autorima je bilo evidentno da oralno zdravlje kod pacijenata s PKU-om zahtijeva detaljnije istraživanje jer je dostupno vrlo malo radova o toj temi, a oni objavljeni radovi pretežno su bili usmjereni na oralno zdravlje djece. Nadalje, u dostupnim istraživanjima izvješćuje o kontradiktornim rezultatima. Na primjer, Kilpatrick i suradnici (12), pregledali su 40 djece s PKU-om i nisu pronašli nikakvu razliku u količini zubnoga karijesa u usporedbi sa zdravom kontrolnom skupinom podudarne dobi/spola. Međutim, znatno više djece s PKU-om imalo je znakove trošenja zuba u usporedbi sa svojim zdravim vršnjacima (33 % prema 24 %). Nedavno, u skladu s našim rezultatima, Ballikaya i suradnici (13), izvijestili su o većoj prevalenciji karijesa među pacijentima s PKU-om. Ipak, treba napomenuti da su u njihovo istraživanje uglavnom bili uključeni vrlo mladi pacijenti (od 1 do 5 godina), a samo mali postotak (16,2 %) bio je stariji od 11 godina. U skladu s našim rezultatima, da Costa Silveira i suradnici (14), otkrili su da je 75 % pacijenata s PKU-om u visokom riziku od nastanka karijesa. Zaključili su da su prevencija i liječenje karijesa veoma važni u stomatološkoj skrbi za te pacijente. Jedno od najopsežnijih istraživanja o toj temi proveli su Singh-Hüsgen i suradnici (15). Ispitali su oralno zdravlje 283 djeteta s PKU-om i otkrili da su imali višu razinu karijesa od onih u zdravoj kontrolnoj skupini, što je sličan nalaz kao u ovom istraživanju za odrasle s PKU-om.

Naše istraživanje pokazalo je da pacijenti s PKU-om imaju smanjenu stopu izlučivanja sline u odnosu prema kontrolnoj skupini. Shimazaki i suradnici (16) ispitali su povezanost brzine protoka sline s prevalencijom zubnoga karijesa i parodontološkim statusom među 2110 odraslih Japanaca te sugerirali da osobe s nižim protokom sline imaju veći rizik od karijesa i parodontitisa. Slina je važan čimbenik u mnoštvu oralnih funkcija kao što su žvakanje, gutanje, antimikrobno djelovanje i djelovanje čišćenja. Slina također utječe na oralno zdravlje kako svojim nespecifičnim fizikalno-kemijskim svojstvima, tako i specifičnim učincima (17).

Važan nalaz u našem istraživanju bio je veći rizik od parodontitisa kod pacijenata s PKU-om. To je u skladu s drugim istraživanjima. Na primjer, Lucas i suradnici (18) pronašli su značajno veći srednji rezultat plaka za 41 pacijenta s PKU-om u usporedbi s kontrolom. Ballikaya i suradnici (13) otkrili su umjereno nakupljanje plaka i upalu gingive kod pacijenata s PKU-om, pri čemu su gotovo svi zahtijevali profesionalne postupke oralne higijene zbog prekomjerne količine plaka i kamenca. Ocjene za nekoliko indeksa oralne higijene bili su primjetno viši za naše pacijente s PKU-om u usporedbi s njihovim kontrolama, što upućuje na povećani rizik od parodontitisa. No treba napomenuti da CPITN indeks ima neka ograničenja jer se temelji na postupnom ocjenjivanju. Nedostaje mjerenje pomičnosti zuba i gubitka pričvrstka, što su važni klinički pokazatelji parodontitisa. Taj indeks ne bi trebao biti jedini parametar za dijagnosticiranje parodontitisa, on je samo alat koji se često upotrebljava u epidemiološkim istraživanjima da bi se utvrdilo je li pacijent u opasnosti od parodontoloških problema (19). Glavni čimbenik rizika za

risk factor for periodontal disease development is a constant presence of dental plaque (20). As expected from previous reports, most of our PKU patients struggled with maintaining good oral hygiene habits, brushing their teeth once a day or only a couple of times a week. Consequently, this is likely to lead to plaque accumulation and, over time, to periodontal disease. Oral hygiene and gingival health were significantly worse for the examined PKU patients compared to the control group, indicating that most PKU patients are at risk of periodontal disease.

Interestingly, there is evidence from previous studies that periodontal disease development can also be influenced by diet. The management of PKU involves adherence to a low-protein diet. It is essential to restrict the intake of Phe to only the amount necessary for normal protein synthesis that can ensure growth and development. Additionally, to make up the energy requirements, their diets are often high in carbohydrates, which are usually taken frequently throughout the day (12). This type of diet is highly cariogenic and potentially erosive. Furthermore, PKU patients often reported snacking every two hours throughout the day, which can also be associated with a higher risk of caries development (12). It has been reported that an exceeded amount of carbohydrates can promote chronic inflammatory diseases (21,22). Moreover, nutritional factors have been implicated in several chronic inflammatory diseases that are associated with periodontitis.

Periodontitis is associated with low micronutrient levels in serum/plasma which may result from dietary and/or lifestyle factors (23). Growing evidence shows that a carbohydrate-rich diet increases the risk of inflammation and gingival bleeding. A diet low in carbohydrates, rich in omega-3 fatty acids, rich in vitamin C and D, and rich in fibre can significantly reduce gingival and periodontal inflammation (24-26). Omega-3 long-chain polyunsaturated fatty acid levels are often reduced for PKU patients, because the primary dietary source of these fatty acids is oily seafood (27,28). Indeed, Bosdet et al. (28), examined 35 adults with PKU (aged 18-46) and found that plasma docosahexaenoic acid (an omega-3 fatty acid found in cold-water, fatty fish)- levels were significantly lower compared to controls.

Despite all the aforementioned oral disease risk factors, our questionnaire revealed that PKU patients visit the dentist significantly less frequently than their healthy counterparts. Prevention of dental caries and periodontal disease depends on the patient's home care measures and regular dental visits. Gingivitis is the only stage of periodontal disease that is reversible, as long as the patient implements and maintains an impeccable oral hygiene routine (29). To avert severe periodontal disease development, it is crucial to implement preventive measures, such as patient information and motivation on how and why to improve their oral hygiene routine (30,31). Patients who visit their dentists frequently are more likely to maintain a good rapport with them and therefore will be better informed and motivated to take care of their oral health. Periodontal disease should be diagnosed and treated as early as possible with professional oral hygiene procedures and periodontal treatment (32). Ultimately, PKU patients should be reminded and motivated by their geneti-

razvoj parodontitisa jest stalna prisutnost zubnog plaka (20). Kao što se i očekivalo na temelju doadašnjih izvješća, većina naših pacijenata s PKU-om imala je problema s održavanjem oralne higijene, pranjem zuba jedanput na dan ili samo nekoliko puta na tjedan. Posljedično, to će vjerojatno rezultirati nakupljanjem plaka i, s vremenom, pojavom parodontitisa. Oralna higijena i zdravlje gingive bili su znatno lošiji kod pacijenata s PKU-om u odnosu prema kontrolnoj skupini, što pokazuje da je većina pacijenata s PKU-om u riziku od parodontitisa.

Zanimljivo je da postoje dokazi u dosadašnjim istraživanjima da na pojavu parodontitisa može utjecati i prehrana. Liječenje PKU-a uključuje prehranu s niskim udjelom proteina. Bitno je ograničiti unos Phe samo na količinu potrebnu za normalnu sintezu proteina koja može osigurati rast i razvoj. Osim toga, kako bi nadoknadili energijske potrebe, njihova prehrana često je bogata ugljikohidratima koji se obično često konzumiraju tijekom dana (12). Ta vrsta prehrane vrlo je kariogena i potencijalno erozivna. Nadalje, pacijenti s PKU-om često su prijavljivali da danju grickaju svaka dva sata, što također može biti povezano s većim rizikom od pojave karijesa (12). Prijavljeno je da prekomjerna količina ugljikohidrata može potaknuti kronične upalne bolesti (21, 22). Štoviše, nutritivni čimbenici uključeni su u nekoliko kroničnih upalnih bolesti koje su povezane s parodontitisom.

Parodontitis je povezan s niskim razinama mikronutrijenata u serumu/plazmi, što može biti posljedica prehrane i/ili načina života (23). Dokazi pokazuju da prehrana bogata ugljikohidratima povećava rizik od upale i krvarenja gingive. Prehrana s malo ugljikohidrata, bogata omega-3 masnim kiselinama, vitaminima C i D te vlaknima može znatno smanjiti upalu gingive i parodonta (24 - 26). Razine dugolančanih polinezasićenih masnih kiselina omega-3 često su smanjene kod pacijenata s PKU-om, jer je primarni izvor tih masnih kiselina masna morska hrana (27,28). Bosdet i suradnici (28), pregledali su 35 odraslih osoba s PKU-om (u dobi od 18 do 46 godina) i otkrili da su razine dokozaheksaenske kiseline u plazmi (omega-3 masne kiseline pronađene u hladnoj vodi, masnoj ribi) bile značajno niže u usporedbi s kontrolom.

Unatoč svim navedenim čimbenicima rizika za oralne bolesti, naš upitnik otkrio je da pacijenti s PKU-om znatno rjeđe posjećuju stomatologa od svojih zdravih parova. Prevencija karijesa i parodontitisa ovisi o mjerama kućne higijene pacijenta i redovitim posjetima stomatologu. Gingivitis je jedini reverzibilni stadij parodontalne bolesti sve dok pacijent primjenjuje i održava besprijekornu oralnu higijenu (29). Da bi se spriječio razvoj teškoga parodontitisa, ključne su preventivne mjere kao što je informiranje pacijenata i motivacija kako i zašto poboljšati oralnu higijenu (30,31). Pacijenti koji često posjećuju stomatologa vjerojatnije će imati dobar odnos s njime i zato će biti bolje informirani i motivirani da se skrbu o svom oralnome zdravlju. Parodontitis treba dijagnosticirati i liječiti što je prije moguće profesionalnim postupcima oralne higijene i parodontološkom terapijom (32). U konačnici, pacijente s PKU-om treba motivirati tijekom svakoga rutinskoga posjeta i podsjetiti koliko su važne pravilne navike kad je riječ o oralnoj higijeni i česti posjeti stomatologu i dentalnom higijeničaru.

cist during every routine visit about the importance of proper oral hygiene habits and frequent visits to the dentist and hygienist.

There has been great speculation concerning the possible reasons for high caries incidence in PKU patients. A priority of parents of children with PKU is maintaining an optimal general health and daily functioning of the child rather than focusing on prevention of oral disease. Children with chronic medical conditions and their families have many pressures placed upon them and there is often a delay in seeking dental care as it is simply not a priority (33). Another important variable that determines the overall wellbeing of PKU patients, including their oral health, is the level of education of the parents. It has been reported that the higher the education level of the parents, the greater the understanding of the disease and the greater the support for the affected child (34). As for adults with PKU, it has been proposed that the increased risk of oral health issues may be due to social burdens. The families of children with PKU face several social issues throughout their childhood, as well as difficulties obtaining the recommended amino acid mixture and providing proper nutrition. In the 1990s, the economic situation in Latvia was poor and the availability of various fruits and vegetables was limited, resulting in increased consumption of starchy products and simple carbohydrates. An increased consumption of such staples has long been determined to be one of the main causes of dental caries and periodontal diseases. Patients who are diagnosed with PKU later in life do not practice a healthy lifestyle, which is optimal for their condition. They have not followed a strict Phe-free diet for years and have not taken the prescribed amino acid formula. Consequently, their general health and mental development are affected and it becomes more difficult to take proper care of their oral health. As detailed in the current article, just over a half of the 45 PKU patients reported brushing their teeth twice a day and only a quarter reported using supplementary dental care products. Previous studies have reported that adult patients feel a sense of guilt about this (35).

It is important for geneticists, dentists and general practitioners to be aware of the findings of this study and previous studies regarding PKU patients and their oral health. Although more research is needed on this subject, it is currently evident that oral health is an issue for PKU patients. Therefore, all medical specialists who encounter PKU patients should recommend early oral health care to spare them future dental costs and the discomfort of toothache and periodontal disease.

Conclusions

Overall, PKU patients have a significantly worse dental and periodontal status than healthy controls. PKU patients in the current study had a higher prevalence of carious teeth than controls. The values of oral hygiene indices (Silness-Löe plaque index, CPITN index and Greene-Vermillion index) were significantly higher in PKU patients compared to controls, indicating a worse oral health status and an increased risk of periodontal disease. PKU patients with the

Uveliko se nagadalo o mogućim razlozima za visoku incidenciju karijesa kod pacijenata s PKU-om. Prioriteti roditelja djece s PKU-om više su u održavanju optimalnoga općega zdravlja i svakodnevnog funkcioniranja djeteta, a ne u prevenciji oralnih bolesti. Djeca s kroničnim zdravstvenim stanjima i njihove obitelji pod mnogim su pritiscima i često se kasno traži stomatološka skrb jer jednostavno nije prioritet (33). Još jedna važna varijabla koja određuje opću dobrobit pacijenata s PKU-om, uključujući njihovo oralno zdravlje, jest stupanj obrazovanja roditelja. Zna se da što je viši stupanj obrazovanja roditelja, to je bolje razumijevanje bolesti i veća je potpora oboljelom djetetu (34). Kada je riječ o odraslima s PKU-om, predloženo je da povećani rizik od problema s oralnim zdravljem može biti posljedica društvenih opterećenja. Obitelji djece s PKU-om tijekom njihova djetinjstva suočavaju se s nekoliko socijalnih problema te s poteškoćama u dobivanju preporučene mješavine aminokiselina i osiguravanju pravilne prehrane. Devedesetih godina prošlog stoljeća ekonomska situacija u Latviji bila je loša, a dostupnost različitoga voća i povrća ograničena, što je rezultiralo povećanom potrošnjom škrobnih proizvoda i jednostavnih ugljikohidrata. Već dugo se zna da je povećana potrošnja takvih osnovnih namirnica jedan od glavnih uzroka karijesa i parodontitisa. Pacijenti kojima se dijagnosticira PKU poslije u životu ne prakticiraju optimalan način života potreban za svoje stanje. Godinama se ne drže stroge dijeta bez Phe i ne uzimaju propisanu formulu aminokiselina. To utječe na njihovo opće zdravlje i mentalni razvoj, što otežava pravilnu brigu o oralnome zdravlju. Kao što je detaljno opisano u ovom članku, nešto više od polovine od 45 pacijenata s PKU-om izjavilo je da pere zube dva puta na dan, a samo četvrtina rekla je da upotrebljava dodatne proizvode za njegu zuba. U dosadašnjim istraživanjima istaknuto je da odrasli pacijenti zbog toga osjećaju krivnju (35).

Važno je da genetičari, stomatolozi i liječnici opće prakse budu svjesni nalaza u ovom istraživanju ali i dosadašnjih istraživanja o pacijentima s PKU-om i njihova oralnoga zdravlja. Iako je potrebno više istraživanja o toj temi, trenutačno je očito da je oralno zdravlje problem za pacijente s PKU-om. Zato bi svi liječnici specijalisti koji rade s pacijentima s PKU-om trebali preporučiti ranu oralnu zdravstvenu zaštitu kako bi ih pošteđjeli budućih stomatoloških troškova i nelagode zbog zubobolje i parodontitisa.

Zaključci

Općenito, pacijenti s PKU-om imaju znatno lošiji dentalni i parodontološki status od onih u zdravoj kontrolnoj skupini. Pacijenti s PKU-om u aktualnom istraživanju imali su veću prevalenciju zuba s karijesom od kontrolne skupine. Vrijednosti indeksa oralne higijene (Silness-Löeov plak indeks, CPITN indeks i Greene-Vermillionov indeks) bile su značajno veće kod pacijenata s PKU-om u odnosu prema kontrolnoj skupini, što upućuje na lošije stanje oralnoga

highest caries prevalence were those who got a delayed PKU diagnosis. PKU patients with the highest caries prevalence showed the least interest in regular visits to the dentist and oral hygienist, according to their questionnaires. In contrast, individuals who were diagnosed with PKU during neonatal screening and followed a low-protein diet properly were more likely to visit the dentist and dental hygienist regularly. Consequently, they had a better dental/gingival health status.

Almost all PKU patients who participated in the current study required professional oral hygiene procedure. Additionally, they predominantly had a decreased basal salivary secretion rate. Evidence shows that PKU patients are at an increased risk of periodontal disease, and they should be informed about it. An effective way of increasing their awareness could be regular reminders from their healthcare professionals that regular dental visits are essential for maintaining optimal general and oral health. A timely diagnosis might be an important prerequisite to reduce the risk of periodontal disease development among PKU patients. PKU patients should visit the dentist regularly in order to ensure early diagnosis of caries and periodontal disease, and the dental hygienist every 3 to 6 months to prevent plaque and calculus build-up. Furthermore, they need to be recommended to rinse their mouth with water immediately after consuming PKU formula to counteract the acidity in their oral cavity.

Conflict of interest

The authors have reported no conflicts of interest.

Author's contribution: I.A. – Study concept and design, analysis and interpretation of data, clinical examination; D.E.E. – Acquisition of data, assistance in clinical examination; I.S. – Critical revision of the manuscript, administrative support; A.B. – Critical revision of the manuscript, recruiting of patients; L.G. – Statistical analysis, administrative, technical support; M.A. – Study supervision, drafting of the manuscript, treatment of PKU patients and recruiting them

zdravlja i povećan rizik od parodontitisa. Pacijenti s PKU-om s najvećom prevalencijom karijesa bili su oni koji su dobili odgođenu dijagnozu PKU-a. Pacijenti s PKU-om s najvećom prevalencijom karijesa, prema podatcima iz upitnika, pokazali su najmanji interes za redovite posjete stomatologu i oralnom higijeničaru. Suprotno tomu, osobe kojima je dijagnosticiran PKU tijekom neonatalnoga probira i pravilno su slijedile dijetu s niskim udjelom proteina, češće su redovito posjećivale stomatologa i dentalnoga higijeničara i posljedično imale bolji zdravstveni status zuba i gingive.

Gotovo svi pacijenti s PKU-om koji su sudjelovali u aktualnom istraživanju zahtijevali su profesionalnu oralnu higijenu. Uz to, pretežno su imali smanjenu bazalnu sekreciju sline. Dokazi pokazuju da su pacijenti s PKU-om izloženi povećanom riziku od parodontitisa te ih o tome treba obavijestiti. Učinkovit način da se poveća njihova svijest o tome može biti redovito podsjećanje liječnika da su redoviti posjeti stomatologu nužni za održavanje optimalnoga općega i oralnoga zdravlja. Pravodobna dijagnoza može biti važan preduvjet za smanjenje rizika od pojave parodontitisa kod pacijenata s PKU-om. Pacijenti s PKU-om trebaju redovito posjećivati stomatologa kako bi se osigurala rana dijagnoza karijesa i parodontitisa, a dentalnog higijeničara svakih 3 do 6 mjeseci kako bi spriječili nakupljanje plaka i kamenca. Nadalje, preporučuje se ispirati usta vodom odmah poslije konzumacije PKU formule da bi se spriječila kiselost u usnoj šupljini.

Sukob interesa

Autori nisu bili u sukobu interesa.

Doprinos autora: I. A. – koncept i dizajn istraživanja, analiza i interpretacija podataka, klinički pregled; D. E. E. – prikupljanje podataka, pomoć pri kliničkom pregledu; I. S. – kritička revizija rukopisa, administrativna potpora; A. B. – kritička revizija rukopisa, odabir pacijenata; L. G. – statistička analiza, administrativna i tehnička potpora; M. A. – nadzor istraživanja, pisanje teksta, liječenje pacijenata s PKU-om.

Sažetak

Uvod: Fenilketonurija (PKU) je autosomno recesivni nasljedni poremećaj metabolizma fenilalanina koji nastaje zbog nedostatka fenilalanin hidroksilaze. Cilj ovog istraživanja bio je procijeniti dentalni status i parodontološko zdravlje pacijenata s PKU-om u Latviji. **Materijal i metode:** Za istraživanje poprečnoga presjeka odabrano je 45 pacijenata s PKU-om i kontrolna skupina podudarne dobi/spola. Njihove anamnestičke podatke, parodontološko zdravlje i dentalni status procijenio je iskusni stomatolog. **Rezultati:** Dentalna i parodontološka klinička procjena pokazala je da je medijan broja zuba s ispunima značajno manji među pacijentima s PKU-om u usporedbi s kontrolnom skupinom ($p = 0,021$). Pacijenti s PKU-om imali su značajno veći medijan broja zuba s karijesom od zdravih ispitanika ($p < 0,001$). Značajne razlike između pacijenata s PKU-om i kontrolne skupine uočene su za nekoliko indeksa oralne higijene ($p < 0,001$): Silness-Löeov indeks plaka, OR = 29,3 (95 % CI: 3,7 – 232,4); CPITN indeks, OR = 35,2 (95% CI: 4,5 – 278,3); Greene-Vermillionov indeks, OR = 10,2 (95 % CI: 2,8 – 38,0), a tu je i potreba za uklanjanjem kamenca, OR = 12,3 (95 % CI: 3,3 – 45,4). **Zaključak:** Utvrđeno je da su dentalni status i parodontološko zdravlje pacijenata s PKU-om znatno lošiji u odnosu prema zdravim ispitanicima u kontrolnoj skupini. To je vjerojatno zbog redovite konzumacije formule za PKU i poteškoća koje imaju mentalno i/ili tjelesno invalidni pacijenti s PKU-om kada je riječ o održavanju oralne higijene. Kako bi spriječili karijes i parodontološke bolesti, profesionalna oralna higijena bila bi pacijentima s PKU-om potrebna svakih tri do šest mjeseci. Nadalje, trebali bi steći naviku ispiranja usta vodom odmah nakon konzumiranja formule za PKU kako bi smanjili kiselost u usnoj šupljini.

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Adresa za dopisivanje

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