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PRIKAZ SLUČAJA
CASE REPORT

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Protetska rehabilitacija pacijenta s keidokranijalnom disostozom: Prikaz slučaja

Prosthetic Rehabilitation of a Patient with Cleidocranial Dysostosis: A Clinical Report

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Sažetak
Kod pacijenata s keidokranijalnom disostozom zahvaćeni su i zubi i čeljust, a karakteristične su pojave prekobrojni zubi, zakašnjela resorpcija mliječnih zuba i erupcija trajnih te impaktirani zubi i možda folikularne ciste. Pacijent u dobi od 26 godina s dijagnozom keidokranijalne disostoze, protetski je bio rehabilitiran. Najprije su bile izrađene privremene pomične proteze sa zadaćom da se procijeni može li se prilagoditi novoodređenoj visini međučeljusnoga odnosa i kako bi se eventualno potaknulo izrastanje trajnih zuba. Naknadno su izrađene teleskopske krunice, korijenska kapica s kuglastim pričvrstkom tipa Dalla Bona i djelomične pomične proteze s metalnom bazom. To je omogućilo zadovoljavajuću funkciju i estetiku, a istodobno je riješen problem izgleda pacijentova lica, povezan s tom bolešću.

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Uvod

Keidokranijalna disostoza je sindrom nepoznate etiologije, a karakterizira ga osnovni trijas: aplazija ili hipoplazija klavikule, zakašnjela osifikacija fontanela i sutura svoda lubanje te nasljedno prenošenje (1). Glava bolesnika je brahiokefalična, s više istaknutim frontalnim, a nešto manje parijetalnim i okcipitalnim izbočenjima. Baza nosa je široka, s depresijom i naglašenom nazo-labijalnom brazdom (2). Bolest također zahvaća zube i čeljusti, pa se javljaju prekobrojni zubi, zakašnjela resorpcija korjenova mliječnih zuba i erupcija trajnih, te impakcija i možda folikularne ciste (2,3).

Taj sindrom prvi je opisao Martin još godine 1765., no ime su mu dali Pierre Marie i Saintom, pa se prema njima ponekad i naziva (4). Tada još nije

Introduction

Cleidocranial dysostosis is a disease of unknown etiology characterized by a triad: clavicular aplasia or hypoplasia, delayed ossification of the fontanelles and sutures of the vault of the skull and hereditary transmission (1). Patient's head is brachiocephalic with expressive frontal prominence, and lesser expressive parietal and occipital prominences. Nose basis is wide with depression and expressive nasolabial furrow (2). Also, it concerns both teeth and jaw and is characterized by supernumerary teeth, delayed resorption of deciduous teeth and delayed eruption of permanent teeth, tooth impaction and possible formation of follicular cysts (2,3).

This syndrome was first described in medical literature by Martin in 1765, but the name for this

bilo rendgenskih zraka te nisu niti mogli potvrditi dva glavna obilježja - aplaziju ili hipoplaziju klavikule i zakašnjelu osifikaciju fontanela (2).

Retencija trajnih zuba i njihova zakašnjela erupcija također su vrlo česte. Najčešće izrastaju prvi trajni molari i donji sjekutići, no može se reći da za izrastanje trajnih zuba uopće ne postoji pravilo. Winter (5) je istaknuo kako trajni zubi bez mliječnih prethodnika imaju veću mogućnost da izrastu. Obično su trajni zubi mikrodontni s kratkim i zakrivljenim korijenima. Prekobrojni zubi obično su slični susjednima, no mogu biti deformirani kukastim korijenima, što može otežati njihovu ekstrakciju. Kod toga sindroma vrlo su česte folikularne ciste, što može završiti velikim defektima kostiju i rezultirati patološkom frakturom (6).

Tijekom posljednjih desetljeća bilo je nekoliko različitih mišljenja o tome što valja učiniti s neizraslim zubima. Najprije se predlagalo da se sve neizrasle zube treba izvaditi, zbog mogućih cista s posljedičnom destrukcijom kosti (6). Kasnije je prihvaćen puno konzervativniji pristup - neizrasle zube ne treba dirati, osim ako se ne počnu stvarati ciste (8). Tako ti neizrasli zubi imaju zadaću sačuvati zaobljen alveolarni greben i smanjiti resorpciju kosti, a time se postiže dobra stabilnost i retencija pomične proteze (9).

Razvojne anomalije trajne denticije osobito su važne za planiranje protetske rehabilitacije pacijenata s cleidocranijalnom dysostozom. Kelly i Nakamoto (8) naveli su zadatke protetske rehabilitacije tijekom zbrinjavanja oralnih manifestacija te bolesti: uspostavljanje vertikalne dimenzije okluzije, protrudiranje (vestibularno dograđivanje) maksile, uspostava funkcionalne okluzije te poboljšanje izgleda i fonacije, a time i poboljšanje mentalnoga zdravlja pacijenta. Može se reći kako estetika sigurno nije manje važna funkcija, čak i kod pacijenata s tako kompleksnom problematikom (10).

Klinički slučaj

Pregledan je bio dvadesetšestogodišnji muškarac s keidokranijalnom disostozom koji je prije toga bio kirurški obrađen. Izvađeni su mu bili svi mliječni zubi i obavljena kortikotomija, kako bi se omogućilo izrastanje trajnih zuba. Zbog gubitka zuba i loših međučeljusnih odnosa, mogućnost žvakanja

syndrome was given by Pierre Marie and Saintom, and therefore it is sometimes named after them (4). They did not dispose of X-rays at that time, so they were not able to confirm two main clinical signs: clavicular aplasia or hypoplasia and delayed ossification of the fontanelles (2).

Retention of the permanent teeth and their delayed eruption is also very frequent. First permanent molars and lower incisors have the greatest eruptive incident, however, it could be stated that the eruption of permanent teeth has no rule. Winter (5) noticed that permanent teeth which had no preliminary deciduous teeth had a greater chance to erupt. Usually permanent teeth are microdontic in whole with short and curved roots. Supernumerary teeth are usually similar to their neighbours but they could be deformed with hook shaped roots causing difficult extractions. In this syndrome the findings of follicular cysts are very often, and that could lead to greater bone destructions causing pathological fractures (6).

During the last decades there have been a few divergent options on what should be done with the unerupted teeth. Firstly, it was suggested that all unerupted teeth should be extracted due to possible cyst formation with subsequent bone destruction (7). Afterwards, more conservative approach was established indicating that unerupted teeth should remain undisturbed unless cyst formation is observed (8). The unerupted teeth serve a purpose in preserving a well-rounded alveolar ridge and in decreasing the bone resorption, thus enhancing the stability and retention of a removable prosthesis (9).

Developmental deformities of the permanent dentition are of primary concern in the prosthetic treatment of patients suffering from cleidocranial dysostosis. Kelly and Nakamoto (8) listed the objectives of prosthetic treatment of the oral manifestations of cleidocranial dysostosis as restorating the vertical dimension of occlusion, building out the maxilla, establishing a functional occlusion, improving appearance and phonation, and improving patient's mental well-being. It could also be said that esthetics is definitely not less important than function even for the patients with such a complex problem (10).

Clinical report

A 26 year old male patient with cleidocranial dysostosis was examined. He had been surgically treated. All deciduous teeth had been extracted, and corticotomy was performed in order to enable eruption of permanent teeth. Chewing ability and esthetics were poor because of missing teeth and

i estetika bili su izrazito loši. Izrasli su desni gornji prvi i drugi molar, lijevi gornji prvi molar, dok je lijevi gornji središnji sjekutić bio tek djelomice izrastao. Zato je na tom zubu obavljen endodontski zahvat, kako bi se omogućila intraradikularna retencija za buduću korjensku kapicu. U donjoj čeljusti izrasli su desni prvi molar, desni drugi premolar i lijevi očnjak, a desni očnjak i desni prvi premolar bili su tek djelomice vidljivi, što je posljedica kortikotomije (Slika 1.).

Najprije su bile izrađene privremene pomične proteze, kako bi se ocijenila sposobnost pacijenta da se prilagodi novoodređenoj vertikalnoj dimenziji okluzije i da se možda potakne izrastanje trajnih zuba (Slika 2.). Planirana vertikalna dimenzija okluzije bila je određena 3 mm od položaja fiziološkoga mirovanja, kako je opisao Posselt (11). Privremene pomične proteze također su imale zadaću povećati žvačnu površinu, odrediti položaj labijalne postave prednjih (umjetnih) zuba te smanjiti emocionalne probleme pacijenta zbog lošeg izgleda lica. Bilo je zadovoljstvo uočiti trenutačno i dojmljivo poboljšanje njegova izgleda. Privremene djelomične proteze nosio je godinu dana. Kako se s njima ugodno osjećao, bilo je odlučeno da i konačnu rehabilitaciju valja obaviti u skladu s njima. Nažalost, retinirani zubi nisu izrasli, a ni oni djelomice izrasli nisu izrasli više. Retinirane zube ne mora se uvijek poticati na izrastanje pritiskom koji čini baza proteze (9). Zbog mnogo impaktiranih zuba nije bilo pogodnih mjesta za postavljanje implantata (usadaka) u čeljusti, pa su zato svi izrasli zubi izbrušeni i uzeti su otisci vinilnim polisiloksanom (Express, 3 M Espe, St. Paul, MN, SAD). Na lijevom gornjem središnjem sjekutiću izrađena je korijenska kapica s intraradikularnom retencijom i kuglastim pričvrstkom tipa Dalla Bona (Servo-dental, Hagen-Halden, Njemačka), a modificirane teleskopske krunice stavljene su na desni gornji prvi i drugi molar, te na lijevi gornji prvi molar (Slika 3.). Prilagođene teleskopske krunice bile su, zbog nerazmjera u međučeljusnim odnosima, izrađene bez faseta i žvačnih ploha, pa su služile samo kao retentivni elementi za djelomične proteze s metalnom bazom (Slika 4. i 5.). Na zubima gornje čeljusti izabrana je ta vrsta konstrukcije kako bi se omogućila dobra retencija i estetika s većom slobodom u prostoru za ispravno postavljanje zuba, te povoljni uvjeti za opterećenje korjenova zuba nosača (12).

„Klasična“ teleskopska krunica bila je izrađena na lijevom donjem očnjaku, također kao retentivni element za donju djelomičnu protezu s metalnom

poor intermaxillary relations. Maxillary right first and second molar and left first molar were erupted, while maxillary left central incisor was just partially erupted. Therefore endodontic therapy was performed on maxillary left central incisor in order to provide intraradicular retention for the future coping. In the mandible right first molar and second premolar were erupted, as well as left canine. Mandible right canine and right first premolar were just slightly visible as a result of corticotomy (Fig. 1).

Firstly, provisional removable partial dentures were made for evaluation of the patient's ability to adapt to the newly determined vertical dimension of occlusion and eventually to stimulate the eruption of permanent teeth (Fig.2). The planned vertical dimension of occlusion was determined to be 3 mm above rest position as described by Posselt (11). Provisional removable partial dentures also served the purpose of increasing the masticatory surface, of determining the labial placement of the anterior artificial teeth, and of alleviating the emotional problems of the patient related to his poor appearance. It was gratifying to notice an immediate and striking improvement in the patient's appearance. The patient wore the provisional removable partial dentures for one year. Since the patient was comfortable with the newly made dentures, it was decided that the final restoration would be constructed accordingly, was no eruption of embedded teeth and slightly erupted teeth did not become more prominent. Embedded teeth are not generally stimulated to erupt by denture bases (9).

Because of the multiple tooth impactions there were no suitable places for implants insertion in the jaws. Therefore, all erupted teeth were prepared and the impressions with vinyl polysiloxane (Express, 3M Espe, St. Paul, MN, USA) were taken. Coping with intraradicular retention with Dalla Bona type stud attachment (Servo-dental, Hagen-Halden, Germany) was made on maxillary left central incisor, and modified telescopic crowns were made for the maxillary right first and second molar, and left first molar (Fig. 3). Due to the discrepancy in intermaxillary relations modified telescopic crowns were made without the veneers and masticatory surfaces, and served just as the retentive elements for metal-base partial denture (Fig. 4, Fig. 5). This construction for maxillary teeth was chosen to provide good retention, esthetics with greater freedom for correct tooth positioning, and more favourable loading conditions for abutment roots (12).

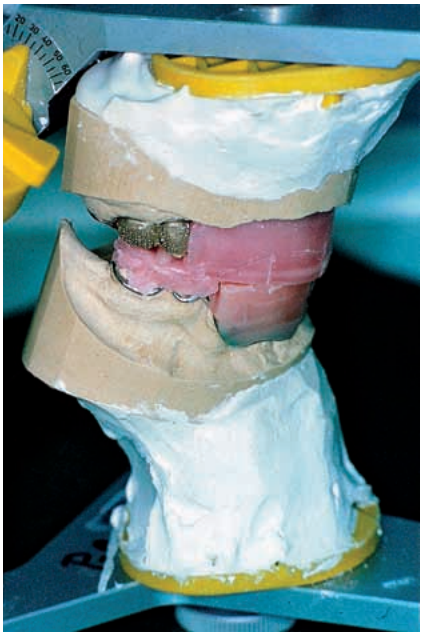
"Classic" telescopic crown was made for the mandibular left canine also as a retentive element for

Slika 1. Oralni status prije protetskog tretmana
Figure 1 Oral status before prosthetic treatment

Slika 2. Postavljanje privremene pomične proteze
Figure 2 Provisional removable dentures in place

Slika 3. Korijenska kapica s pričvrstkom tipa Dalla Bona i unutrašnjim teleskopima
Figure 3 Root coping with Dalla Bona attachment and inner telescopes

Slika 4. Metalna baza pomičnih djelomičnih proteza sa zalemljenim vanjskim teleskopima
Figure 4 Metal-bases of removable partial dentures with soldered outer telescopes



Slika 5. Modeli sa zagriznim šablonama postavljeni u artikulator
Figure 5 Models with bite registration mounted in articulator

Slika 6. Rekonstruktivne proteze u okluziji
Figure 6 Reconstructive dentures in occlusion

bazom, čije su kvačice postavljene na desnom donjem prvom molaru i drugom premolaru - oni su bili intaktni, bez ikakva karijesa (Slika 4.), pa nisu bili opskrbljeni protetskim radom. Kako bi se omogućilo izrastanje donjega desnog očnjaka, bio je izdubljen akrilat na bazi nove proteze.

Svi metalni dijelovi fiksnih protetskih radova bili su od zlatno-platinske legure (18+8, Rafinerija plemenitih kovina, Zagreb, Hrvatska), a metalne baze proteza bile su od legure Co-Cr (Remanium GM 380, Dentaurum, Pfortzheim, Njemačka).

Unatoč deformitetu i nepovoljnim međučeljusnim odnosima, konačne su proteze imale dobru okluziju i estetiku (Slika 6.) te su smanjile deformitete pacijentova lica.

Zaključak

Ne postoji specifičan tretman za abnormalnosti koštanoga tkiva u slučaju cleidocranijalne dysostoze. Protetska rehabilitacija oralnih manifestacija u slučaju te bolesti vrlo je važna, ne samo kako bi se uspostavila oralna funkcija i estetika, nego i riješili ili umanjili problemi povezani s izgledom lica tih pacijenata, a usko su povezani s bolešću. Tijekom planiranja protetskog zahvata, osobito ako nisu uporabljeni usadci - implantati, terapeut mora uzeti u obzir sve izrasle zube i nepovoljne međučeljusne odnose, jer će samo dobra terapija rezultirati zadovoljavajućom funkcijom i estetikom.

Zahvala

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metal-base partial denture the clasps of which were placed on mandibular right first molar and second premolar that were intact, without any decay (Fig. 4), so they were not covered with any prosthetic appliance. Provision for further possible eruption of mandible right canine and right first premolar was made by relieving the resin in the new denture base.

All metal parts of fixed prosthetic appliances were cast in gold-platinum alloy (18+8, Precious Metals Refinery, Zagreb, Croatia), and the metal bases of the dentures were made from Co-Cr alloy (Remanium GM 380, Dentaurum, Pforzheim, Germany). Despite the deformities and unfavourable intermaxillary relations, the final prostheses had satisfactory occlusion and esthetics (Fig. 6) and ameliorated the patient's facial deformities.

Conclusion

There is no specific treatment for the skeletal abnormalities that characterize cleidocranial dysostosis. Prosthetic treatment of the oral condition of patients suffering from cleidocranial dysostosis is very important, not only to restore oral function and esthetics but also to resolve or ameliorate the problems of facial appearance of such patients that are associated with this disease. During prosthetic treatment planning, especially if implants are not used, one should take into consideration all the erupted teeth and the unfavorable intermaxillary relations, because only well-planned therapy will result in satisfactory function and esthetics.

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Abstract
Cleidocranial dysostosis (ccd) concerns both teeth and jaw and is characterized by supernumerary teeth, delayed resorption of deciduous teeth and delayed eruption of permanent teeth, tooth impaction and possible follicular cysts formation. A 26 years old patient with ccd received prosthetic treatment. Firstly, provisional removable prostheses were made for evaluation of the patient's ability to adapt to the newly determined vertical dimension of occlusion and to stimulate the eruption of permanent teeth. Afterwards, telescopic crowns, coping with intraradicular retention and Dalla Bona type stud attachment, and metal-base partial dentures were designed resulting in satisfactory function and esthetics, also resolving the problems of appearance associated with this disease.

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