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A MULTIDISCIPLINARY TREATMENT OF PATIENTS WITH CRANIOFACIAL DISORDERS. OWN EXPERIENCE

SKOJARZONE LECZENIE ORTODONTYCZNO-PROTETYCZNE U PACJENTÓW Z ZABURZENIAMI ROZWOJOWYMI W OBRĘBIE CZĘŚCI TWARZOWEJ CZASZKI. DOŚWIADCZENIA WŁASNE

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Abstract

Oral rehabilitation of patients with craniofacial disorders is a great challenge and needs a multidisciplinary approach. This is due to the diverse etiology of the disease and severity of changes in tissues and organs. Congenital absence of tooth germs manifested in the form of oligodontia or anodontia, the presence of persistent deciduous teeth in ectodermal dysplasia (ED), cleft lip and hard palate or cancer-induced changes in the tissues of the stomatognathic system are the most common causes of these disorders. The observed abnormalities are responsible for functional disorders of musculo – articular system, speech and chewing. In addition, noticeable adverse changes in the appearance have a huge psychological impact on patients and their well-being. Therefore, the treatment of these medical conditions should begin in childhood and comprise interdisciplinary rehabilitation, involving prosthetics and orthodontics supported by surgery, as well as speech or laryngological therapy. In this paper the interdisciplinary treatment of patients with oral hard and soft tissues disorders during ectodermal dysplasia is discussed. Early oral rehabilitation can restore lost or abnormally shaped tissues and proper functions of the masticatory system. It can also have a positive impact on further physical and psychological development of patients.

Key words: ectodermal dysplasia, adolescent, interdisciplinary care

Streszczenie

Rehabilitacja układu stomatognatycznego u pacjentów z zaburzeniami rozwojowymi w obrębie części twarzowej czaszki jest leczeniem skomplikowanym i wymagającym podejścia interdyscyplinarnego. Dzieje się tak ze względu na różną etiopatogenezę oraz stopień nasilenia zmian w obrębie tkanek i narządów. Najczęściej spotykanymi przyczynami zaburzeń jest wrodzony brak zawiązków zębów w postaci oligodoncji lub anodoncji oraz obecność przetrwałych zębów mlecznych w przebiegu dysplazji ektodermalnej oraz rozszczep wargi i podniebienia twardego lub zmiany w obrębie tkanek układu stomatognatycznego spowodowane procesem nowotworowym. Występujące nieprawidłowości są przyczyną zaburzeń czynnościowych układu mięśniowo-stawowego, zaburzeń mowy i żucia. Dodatkowo zauważalne negatywne zmiany w wyglądzie mają ogromny wpływ na psychikę pacjentów i ich samopoczucie. Z tego powodu leczenie powinno rozpoczynać się w okresie dzieciństwa i uwzględniać rehabilitację interdyscyplinarną, ortodontyczno-protetyczną, wspomaganą leczeniem chirurgicznym, logopedycznym czy laryngologicznym. W pracy przedstawiono interdyscyplinarne leczenie pacjentów z zaburzeniami rozwojowymi układu stomatognatycznego w przebiegu zespołu ektodermalnego. Wczesna rehabilitacja pozwala odbudować utracone lub nieprawidłowo wykształcone tkanki, przywracając prawidłową funkcję narządu żucia oraz pozytywnie wpływając na dalszy rozwój fizyczny i psychoemocjonalny pacjentów.

Słowa kluczowe: dysplazja ektodermalna, pacjenci młodociani, leczenie interdyscyplinarne

INTRODUCTION

Teeth loss caused by greatly damaged hard tissues, resulting from carious processes and traumatic dental injuries are the major reasons for prosthetic rehabilitation in children and youth. Cancer outcomes in the craniofacial region and treatment-related complications also lead to disorders of the stomatognathic system. Developmental malformations are another cause of the oral cavity disorders [1, 2]. The most common congenital defects include cleft lip and palate, as well as defects with coexisting loss of teeth, like ectodermal dysplasia (ED) induced by tissue or organ hypoplasia originated from one of the three primary germ layers – the ectoderm. Congenital tissue hypoplasia of the ectodermal origin is a rare congenital defect (incidence, 1:10 000-1:100 000 live births). Clinical symptoms of ectodermal dysplasia are manifested by changes in the skin, mucous membrane, hair, nails and teeth. The patients show pale, thin and dry skin with diminished volume of panniculus adiposus, deficiency of sebaceous glands and excessive keratosis on palms and feet [3]. Two clinical types of this disease are distinguished, hydrotic with normal function and number of sweat glands and hypohydrotic with coexisting disorders manifested by the dry skin and mucous membranes, as well as by decreased lacrimation. Both types are genetically determined, the former is dominantly inherited, the later is recessively inherited and masculine gender-linked. In facial features of ED patients attention is specially drawn by a saddle nose, frontal eminence, retraction of the middle face, low-placed ear concha, sparse and light hair, sparse eyebrows and eyelashes, shortened lower part of the face with strongly marked mentolabial sulcus [2, 3, 4].

Congenital absence of tooth germs in the form of hypodontia, oligodontia, anodontia or rarely aplasia is the major abnormality in the stomatognathic system. Persistent deciduous teeth, disorders in the mineralization of enamel and dentin, impaired shape and localization of teeth in the arch, defective alveolar ridges in the edentulous fragments, lower occlusion height and glossal dysfunction are usually observed. All the aforesaid abnormalities also affect the function of the musculo-articular system, leading finally to speech and mastication disorders, persistent infantile type of swallowing and mouth breathing [2, 3, 5]. Extra- and intraoral disorders exert adverse effects on the patient's face appearance, hence the source of frequent complexes with their negative impact on interpersonal contacts and the cause of rejection by peers or the people around. Therefore a multidisciplinary treatment and holistic approach to the patient, specially relevant to a child, has been discussed in the context to bioethics in Developmental Period Medicine [5].

In this paper two cases of the interdisciplinary treatment of patients with ectodermal dysplasia, using overdentures (OVD) supported by telescopic crowns are discussed.

CASE REPORT 1

A 23-year old patient (S.M.) presented for prosthetic consultation and treatment. Over a number of years the

patient had been under dental care of an orthodontist due to diagnosed ectodermal dysplasia with coexisting mastication disorders. On general examination mild disorders in the facial features and the structure of skin, hair and nails, characteristic of ED patients, were observed. On the intraoral examination significant disproportion of the maxillary and mandibular dental arches, as well as hypoplasia of the mandibular alveolar area in the edentulous segment attracted particular attention (fig. 1). Based on the clinical examination and the analysis of orthopantomographics (OPG), the presence of persistent deciduous teeth in the maxilla and mandible and a few permanent teeth were found (11, 16, 21, 26, 46). Figure 2 presents the dental status of the patient.

The examination also revealed disorders of enamel and dentin mineralization in the whole range of teeth, abnormal shape and dimension of clinical crowns, as well as replacement of maxillary incisors relative to the body medial line. On account of oligodontia and the absence of supporting zones, the occlusal height was significantly diminished.

The preparation of prosthetic treatment began with an interdisciplinary approach to the oral cavity sanitation, conservative and periodontal treatments with a detailed oral hygiene instructions. In stage one of the prosthetic treatment two complete overdentures, upper and lower, were made in constructive occlusion, which covered microdentic clinical crowns (fig. 3). Due to the extent of the upper denture plate, difficulty in the adaptation to prosthetic restorations and young age of the patient, it was



Fig. 1. Intraoral view. The dental status of the patient (S.M.), aged 23, before treatment.

Ryc. 1. Zdjęcie wewnątrzustne – stan uzębienia pacjenta (S.M.), lat 23, przed leczeniem.

6 V IV III 1	1 IV 6
6 V IV III I	I III IV V 6

Fig. 2. Graph shows the dental status of a 23-year old patient (S.M.).

Ryc. 2. Diagram przedstawia status zębowy pacjenta I. 23 (S.M.).

decided to take an additional examination – cone-beam computed tomography (CBCT) and an implanto-prosthetic consultation to consider the treatment based on endosseous implants. Bearing in mind morphological abnormalities within the osseous structures of the stomatognathic system, insufficient bone volume and significant disproportion of dental arches (maxillary micrognathia), the dental implant treatment was finally given up.

Following a thorough analysis of the clinical status and the assessment of a long-term prognosis of maxillary deciduous teeth (taking into account a possible abutment loss), in stage two of the prosthetic treatment the manufacture of the upper denture, supported by telescopic crowns with substantially reduced plate, was planned. The patient was referred to the endodontic treatment of all teeth in maxilla and then abutment teeth were provided with fiber-glass reinforced composite (FRC) posts. This was followed by planning the manufacture

of primary cylindrical telescopic crowns of zirconium oxide. However, due to complications observed in the endodontic treatment of teeth 26 and 53, the idea of their use as prosthetic abutments failed. Therefore, it was decided to leave these two teeth in the oral cavity. Prosthetic abutments were prepared and impressions taken according to commonly adopted principles. On control examination of primary crowns in the patient's mouth gingivitis and unsatisfactory oral hygiene were evidenced. The instruction in oral hygiene principles was repeated and after healing of gingivitis and improving hygiene habits primary crowns were cemented (fig. 4). Teeth 26 and 56 provided additional retention for the complete overdenture with substantially reduced plate, supported by telescopic crowns (fig. 5). Planning the best possible prosthetic solutions and prosthetic constructions, the patient's age, oral cavity clinical status, hygiene habits, prosthetic plate range, restoration esthetics and long-term



Fig. 3. Restorations made in stage one of prosthodontic treatment.

Ryc. 3. Uzupełnienia protetyczne wykonane w I etapie leczenia protetycznego.

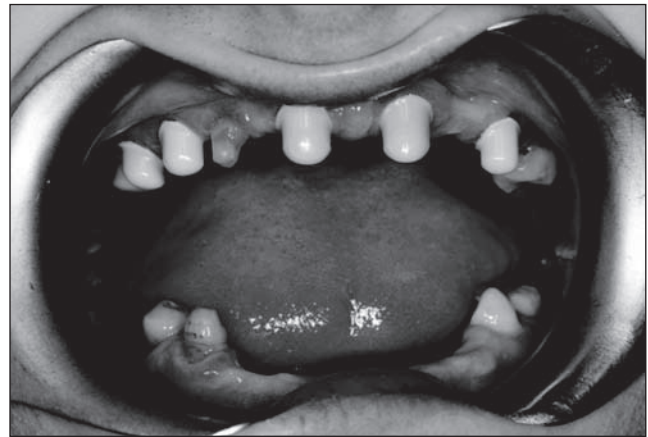


Fig. 4. Cemented internal cylinder crowns made of zirconia dioxide, using CAD/CAM technology.

Ryc. 4. Zacementowane cylindryczne korony wewnętrzne wykonane z tlenku cyrkonu metodą CAD/CAM.

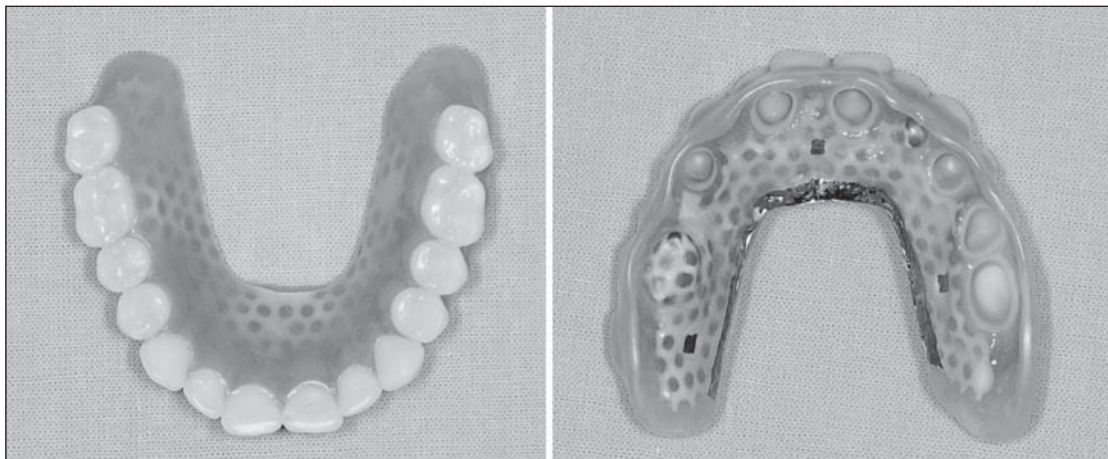


Fig. 5. Complete upper overdenture with significantly reduced plate supported by telescopic crowns and PMMA inserts placed in the external crowns.

Ryc. 5. Proteza całkowita górna typu overdenture ze znacznie zredukowaną płytą wsparta na koronach teleskopowych oraz umieszczonymi wkładkami PMMA w koronach zewnętrznych.



Fig. 6. Extraoral view. Patient (S.M.) with diagnosed ectodermal dysplasia, facial features before and after prosthodontic treatment.

Ryc. 6. Zdjęcie zewnętrzne – pacjent (S.M.) z rozpoznaną dysplazją ektodermalną, rysy twarzy przed leczeniem oraz po zakończonym leczeniu protetycznym.



Fig. 7. Patient (P.K), aged 25, with Ellis-van Creveld syndrome, visible malformations of palms and feet.

Ryc. 7. Pacjent (P.K.), lat 25 z rozpoznanym zespołem Ellisa-van Crevelda- widoczne malformacje dłoni i stóp.

prognosis of the abutment, as well as possible corrections with no need to exchange restorations, were taken into consideration. Figure 6 shows the facial features before and after the prosthodontic treatment.

CASE REPORT 2

A 25-year old patient (P.K.) with diagnosed Ellis-van Creveld syndrome (ectodermal dysplasia, group A, subgroups 1-2-3 according to Freire-Maia and Pinheiro), had been under care of an orthodontist over many years. On extraoral examination sparse hair, pale and thin skin, dropped lower facial segment and more pronounced lower mentolabial sulcus were observed. In addition, symptoms of ED syndrome associated with developmental disorders of other germ layers, such as anomalies of the osseous system, including malformation of palms and

feet, were found (fig. 7). The clinical examination and dental radiographs revealed the presence of maxillary and mandibular permanent molars and maxillary medial incisors. Persistent deciduous teeth, 52 and 53, showed physiological mobility and relatively long non-resorbed roots. Figure 8 presents the dental status of the patient.

6 V IV III II 1	1 IV V 6
6 V IV III III I	I III IV 6

Fig. 8. Graph shows the dental status of a 25-year old patient (P.K.).

Ryc. 8. Diagram przedstawia status zębowy pacjenta l. 25 (P.K.).

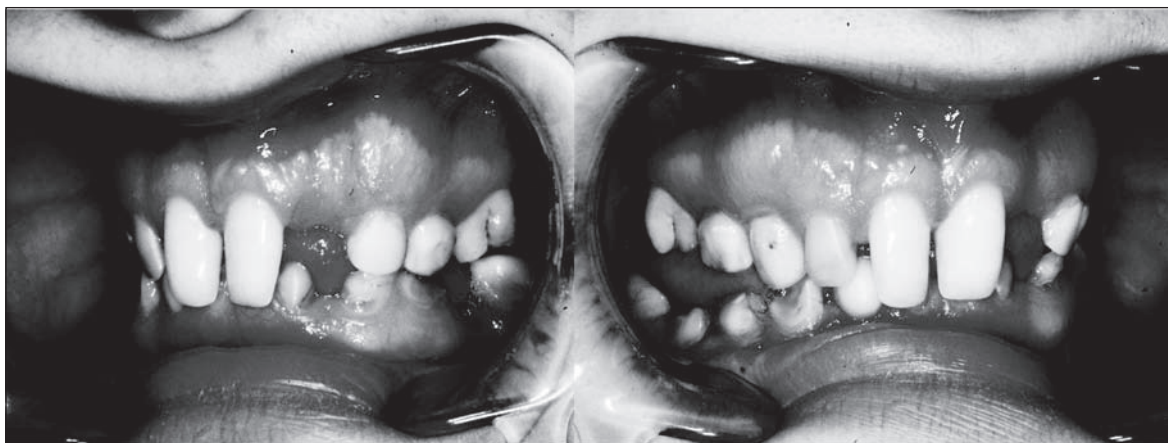


Fig. 9. Intraoral view. The dental status of the patient (P.K.), aged 25, before treatment.

Ryc. 9. Zdjęcie wewnątrzustne – stan uzębienia pacjenta (P.K.), lat 25, przed leczeniem.



Fig. 10. Entering dental implants in the mandible.

Ryc. 10. Wprowadzanie wszczepów śródkostnych w obrębie żuchwy.

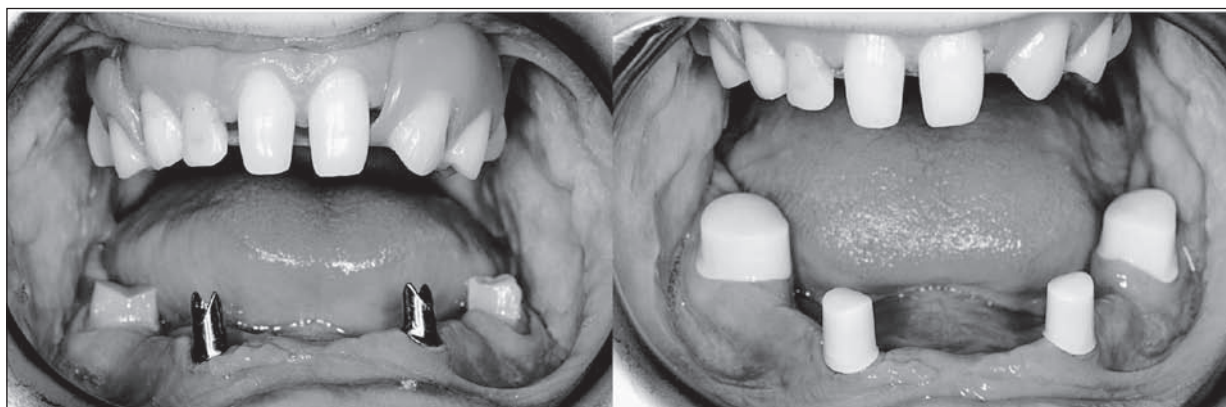


Fig. 11. Embedded implant abutments on implants in the position of mandibular canines and primary crowns of zirconium oxide on prosthetic abutments.

Ryc. 11. Osadzone łączniki implantologiczne na wszczepach w pozycji kłów w żuchwie i korony pierwotne z tlenku cyrkonu na filarach protetycznych.

Due to significant root resorption, mandibular persistent deciduous teeth were classified to be extracted. On intraoral examination the attention was drawn to the central incisors deeply overlapping and open occlusion in the right lateral segment (fig. 9). In the developmental phase and pubertal spurt the orthodontic treatment was focused on promoting vertical growth of the maxillary alveolar ridges, the alveolar part of the mandible and protruded growth of the mandible with use of Schwartz's plate. In stage two of the orthodontic treatment a block apparatus with constructive occlusion was manufactured.

In phase one of the prosthetic treatment two partial dentures, settled in constructive occlusion, were made. On account of the unsatisfactory lower denture retention the CBCT examination of the mandible was performed and implant-prosthetic consultation was provided. After a thorough analysis of the bone structure, implants in the place of missing teeth, 33 and 43, an overdenture supported by telescopic crowns, covering the patient's own teeth and based on implants, were planned. The patient was referred to the endodontic treatment of tooth

46 and revision of the endodontic treatment of tooth 36. The teeth were supplied with standard FRC posts. The intra-procedure picture shows the entering of dental implants in the place of missing teeth, 33 and 43 (fig. 10). During the period of healing and osseous integration no complications were observed. The achieved proper osteointegration allowed for the fixation of abutments on implants (fig. 11) and continuation of the treatment, according to well known principles. After setting primary crowns made of zirconium dioxide (fig. 11), overdenture was produced. It was reinforced with metal cast construction with placed external crowns with fixed inserts of polymethylmethacrylate (PMMA) (fig. 12). The treatment using permanent maxillary prosthetic restorations will present a consecutive phase of prosthetic rehabilitation (fig. 13).

Owing to prosthetic rehabilitation the retention of the lower denture was significantly improved, making a great impact on its function, mastication and well being of the patient. The effect of replacing missing teeth in the mandible was reflected in proper proportions and

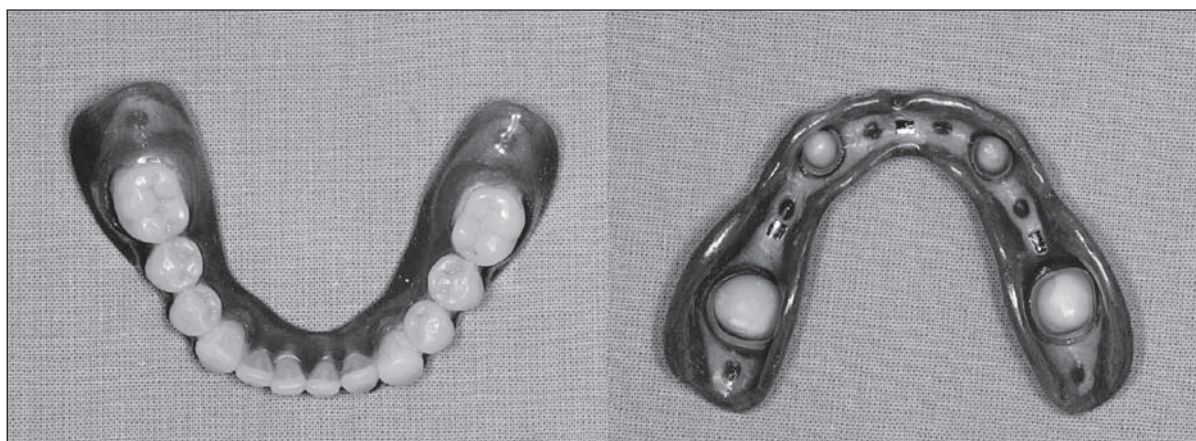


Fig. 12. Lower overdenture with PMMA inserts embedded in external crowns.

Ryc. 12. Dolna proteza typu overdenture z osadzonymi wkładkami PMMA w koronach zewnętrznych.



Fig. 13. Intraoral view. Condition before treatment of the upper jaw and after mandibular prosthetic rehabilitation.

Ryc. 13. Zdjęcie wewnątrzustne – stan przed leczeniem w szczęce i po leczeniu protetycznym w żuchwie.

improved facial appearance of the patient. The patient was informed about the need to continue prosthetic treatment of the upper dental arch and to pay frequent control visits.

DISCUSSION

Stomatognathic system abnormalities occurring in ectodermal dysplasia are responsible for functional disorders of the musculo-articular system, speech and mastication [1, 4-7]. In addition, noticeable adverse changes in the appearance exert a tremendous impact on the mental status and well being of ED patients [1, 6]. When the process of osseous development is completed it is necessary to undertake the next stage of an interdisciplinary rehabilitation of the masticatory organ provided by a team of different specialists [8, 9]. The treatment should be planned individually, depending on the osseous foundation status and the number of missing tooth germs. The treatment can involve fixed or removable prosthetic dentures, blocked crowns and bridges and overdentures supported on own teeth or intraosseous implants [7-11]. Telescopic crowns find application in the case of residual dentition or oligodontia in clinical conditions when muco-osseous foundation relief is required. The use of restorations supported on telescopic crowns improves the access to hygiene procedures, they also play role of splinting and relieving the muco-osseous foundation and in the loss of defective abutments facilitates the correction of restorations with no need for their exchange [12]. In this paper we present an example of using crown-sleeve-coping produced of zirconium oxide in the CAD/CAM technology and secondary with PMMA inserts in denture base. Application of PMMA material allows for improving the retention of restorations after diminished fraction. In addition, a digital record facilitates the repeated production of secondary crowns with no need to take new impressions in difficult clinical prosthetic procedures. Due to application of telescopic crowns it is much easier and faster to repair the restoration if any of the abutments have been lost.

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