

Bogumił Lewandowski[1,2], Katarzyna Martula-Gala[3], Robert Brodowski[1], Barbara Zych[4]

MULTIPLE, SUPERNUMERARY RETAINED TEETH IN THE COURSE OF CLEIDO-CRANIAL DYSPLASIA. A CASE REPORT

MNOGIE, DODATKOWE ZĘBY ZATRZYMANE W PRZEBIEGU DYSPLAZJI CZASZKOWO-OBOJCZYKOWEJ. OPIS PRZYPADKU

[1]Clinical Department of Maxillo-Facial Surgery, Provincial Specialist Hospital in Rzeszów, Poland
[2]Institute of Emergency Medicine, Faculty of Medicine, the University of Rzeszów, Poland
[3]"Primadent", Non-public Health Care Institution, Rzeszów, Poland
[4]Institute of Obstetrics, Faculty of Medicine, the University of Rzeszów, Poland

Abstract

Cleido-cranial dysplasia, often referred to as Scheuthauer-Marie-Sainton syndrome, is an autosomal dominant disorder of the musculo-skeletal system.

Patients with cleido-cranial dysplasia are characterized by short stature, frequent varus or valgus hip, kyphoscoliosis, underdevelopment of the scapulas and the sternum, incorrect number of ribs. The most characteristic feature is unilateral or bilateral, partial or total underdevelopment of clavicles. Mental development is not affected in this syndrome. Malocclusion, occlusal irregularities, multiple supernumerary teeth, impacted teeth, and persistent milk teeth are found in the stomatognathic system. Teeth often have abnormal anatomy. Gothic palate, cleft hard and soft palate are diagnosed.

The aim of this paper is to present a case of a 12-year-old boy diagnosed with irregularities in the masticatory system involving an additional number of retained teeth. The boy was referred by an orthodontist for surgical and orthodontic team therapy. The case presented confirms the observations of other authors that only the multi-specialty collaboration of a pediatrician, a geneticist, an orthopedist, an orthodontist, a maxillofacial surgeon, an implant prosthetic surgeon and a physiotherapist can provide proper diagnosis and treatment.

Key words: cleidocranial dysplasia, supernumerary teeth, impacted teeth, radiological examination

Streszczenie

Dysplazja czaszkowo-obojczykowa zwana zespołem Mariego-Saintona lub zespołem Scheuthauera jest genetycznie uwarunkowaną wadą rozwojową układu kostno-szkieletowego, dziedziczoną w sposób autosomalny dominujący.

Pacjenci w dysplazją obojczykowo-czaszkową charakteryzują się niższą wysokością ciała, często występuje szpotawość lub koślawość stawów biodrowych, kifoskolioza, niedorozwój łopatek i mostka, nieprawidłowa liczba żeber. Najbardziej charakterystyczną cechą jest jedno – lub obustronny, częściowy lub całkowity niedorozwój obojczyków. W zespole tym nie obserwowano zaburzeń rozwoju umysłowo-psychocznego. W narządzie żucia opisywano wady zgryzu, nieprawidłowości zgryzowo-zębowe, mnogie zęby nadliczbowe, zęby zatrzymane oraz przetrwałe zęby mleczne. Zęby często mają nieprawidłową budowę anatomiczną. Stwierdza się podniebienie gotyckie, Roszczep podniebienia twardego i miękkiego.

Celem niniejszej pracy jest prezentacja 12-letniego chłopca, u którego stwierdzono nieprawidłowości w układzie narządu żucia polegające na występowaniu licznych dodatkowych zębów zatrzymanych.

Chłopca skierował ortodonta celem zespołowego leczenia chirurgiczno-ortodontycznego. Prezentowany przypadek własny potwierdza poglądy innych autorów, że tylko wielospecjalistyczna współpraca pediatry, genetyka, ortopedy, ortodonty i chirurga szczękowego, implanto-protetyka oraz rehabilitanta może zapewnić prawidłowe rozpoznanie i leczenie.

Słowa kluczowe: dysplazja obojczykowo-czaszkowa, zęby nadliczbowe, zęby zatrzymane, badanie radiologiczne

DEV PERIOD MED. 2015;XIX,4:503-507

INTRODUCTION

Some genetic conditions are characterized by developmental disorders in the form of multiorgan failure syndromes, including problems in the stomatognathic system and facial bones, as in the case of cleidocranial dysplasia.

Cleidocranial dysplasia (CCD), also known as Scheuthauer-Marie-Sainton syndrome [1, 2], is a rare (1: 1 000 000) congenital disorder of the skeletal system not related to gender and race. It is an autosomal dominant disease and about 1/3 of the cases are new mutations [3]. Mutation affects the RUNX2 / (CBFA1) / gene located on the 6p21 chromosome responsible for the formation of bone structures, the differentiation of osteoblast from precursor cells and chondrocytes' maturation. As a result of the mutation, abnormal osteoblast function and bone disorders occur. There has been a variety of clinical symptoms which are associated with a different expression of the gene / CBFA1 /. Patients affected by this defect have normal intellectual development [4].

The typical CCD clinical symptoms are irregularities in the skeletal system characterized by short stature, which occurs between 4 and 8 years of age [5]. They have an impaired structure of the pelvis associated with the expansion of the pubic symphysis. Pelvic dysplasia in pregnant women in the course of CCD often requires a Caesarean Section. Varus or valgus hip, kyphoscoliosis, joint hypermobility and muscle laxity are observed. Hypoplasia of the distal phalanges of the hand (brachydactyly) can occur in patients with dysplasia [6]. Developmental disorders in nails include their lack or dysplasia [2]. Symptoms in the chest are characterized by irregularities in the build and deformation of the scapulas and the sternum. Anomalies occur in the number of ribs (aplasia or additional cervical ribs), and hypoplasia or aplasia of one or both clavicles [3, 5]. The chest may be tapered or cone-shaped (a bell), which can potentially lead to respiratory failure in early childhood [5, 6].

In terms of skull abnormalities related to its anatomy and shape: i.e. brachycephaly, prominent frontal and occipital bony lumps [2], there may be hypertelorism (wide spacing of sockets), large foramen magnum [6], characteristic delayed obliteration of the cranial and parietal sutures [1, 2, 4] and the presence of multiple Wormian bones [5, 6]. Aplasia or hypoplasia of the nose bone causes the bridge of the nose to becomes flattened and lowered and wide at the base [7]. Partial or no paranasal sinuses are found. In 40% of the cases there is partial conductive hearing loss, which is the result of ossicle deformation. Moreover, the lachrymal, sphenoid, zygomatic and mastoid bones hypoplasia occur [8, 9, 10].

Face proportions in young children remain normal. Later, hypoplasia of the middle part of the face occurs and the chin becomes more protuberant [11]. Disproportion between the maxilla and the mandible and occlusion disorders occur in the form of pseudo protrusive occlusion. Gothic palate, cleft hard and soft palate, delayed or no mandibular symphysis ossification are found in intraoral examination [11].

CCD patients are diagnosed most commonly with abnormal clavicles: bilateral hypoplasia of the shoulder, the middle or the central part of the clavicle [1, 2, 3, 4, 5]. The missing section is replaced by the fibrous tissue and pseudoarthrosis is created [1, 2, 4]. Clavicular aplasia occurs in 10% of the patients [1, 8]. They can bring together the shoulders to meet in front of the chest. CCD patients show no physical development impairment [6].

Apart from craniofacial abnormalities, subjects with Scheuthauer-Marie-Sainton syndrome are also diagnosed with disorders of the teeth and the stomatognathic system. These include delayed replacement of primary teeth by permanent teeth, retained deciduous teeth, supernumerary teeth and impacted teeth around which dentigerous cysts frequently arise [10]. Teeth often have an abnormal anatomical structure [7, 8, 9, 10].

AIM

The aim of this paper is to present the case of a 12-year-old boy diagnosed with CCD recently treated at the Clinical Department of Maxillo-Facial Surgery, at the Provincial Specialist Hospital in Rzeszów.

CASE REPORT

A 12-year-old boy was referred by an orthodontist to the Hospital Maxillofacial Surgery Clinic in Rzeszów for surgical treatment of abnormal teeth, malocclusion and impacted teeth. In the mother's opinion her son requires constant pediatric observation. Dental abnormalities were detected incidentally on a pantomogram during orthodontic consultation, for which the boy was referred by a pediatrician.

A slight facial asymmetry with a prominent frontal bony lump, concave profile of the mandible with a prominent chin and a deeper mentolabial sulcus were found during the extraoral examination. The middle section of the face was shortened with a marked hypoplasia and underdevelopment of the mandible. The nasal bridge was withdrawn with a high nasopharyngeal angle. The nose was unobstructed without pathologic secretions. The eyeballs were properly seated without abnormalities of motion and visual acuity (fig. 1).

The disproportion in the development of the maxilla and the mandible with a tendency to anterior occlusion was found in the extraoral examination. A high arched, narrow so-called "gothic" palate was observed. Numerous tooth abnormalities were found, such as: delayed eruption of permanent teeth and resorption of deciduous tooth roots, persistent deciduous teeth. Gaps were present in permanent teeth. The tooth diagram was as follows:

Tooth diagram:

-, -, 16, 55, - , - , - , 11, 21, - , - , - , 65, 26, - , -

-, -, 46, 85, 84, -, 42, 41, 31, 32, - , 74, 75, 36, - , -

During general examination of other organs, short stature, as well as chest and pelvis deformation were found. Abnormalities in the chest concerned its asymmetry, deformation, lack of clavicles and a collapsed sternum (fig. 2). The chest X-ray showed clavicular aplasia and

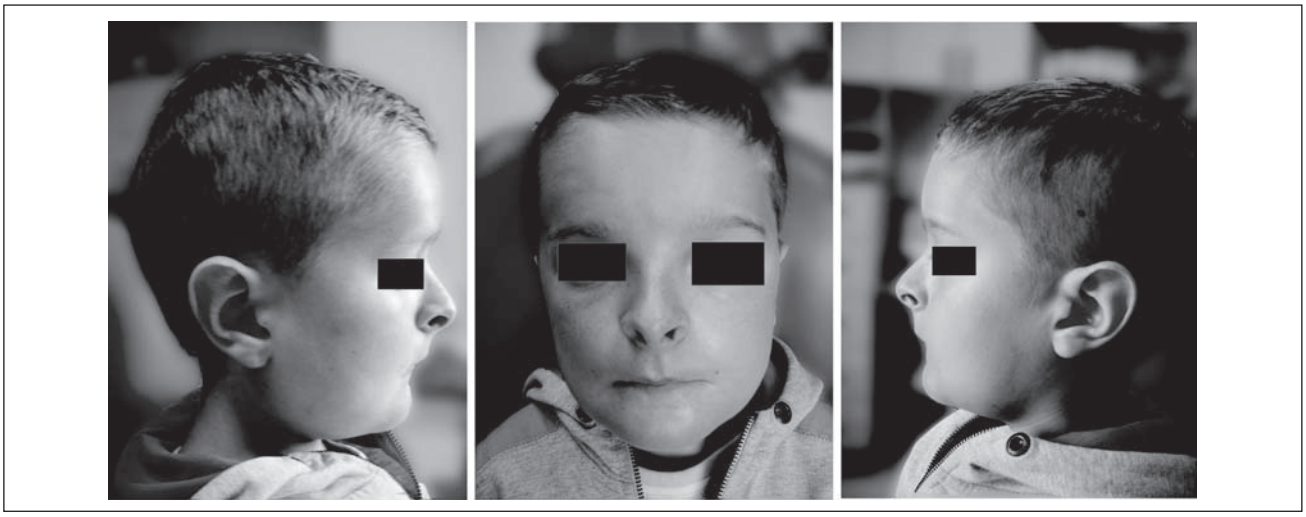

Fig. 1. En face and profile picture of the boy.
Ryc. 1. Zdjęcie chłopca en face i z profilu.

Fig. 2. The photo shows the symptom of „close relations of shoulders".
Ryc. 2. Fotografia obrazująca objaw „zbliżania ramion".

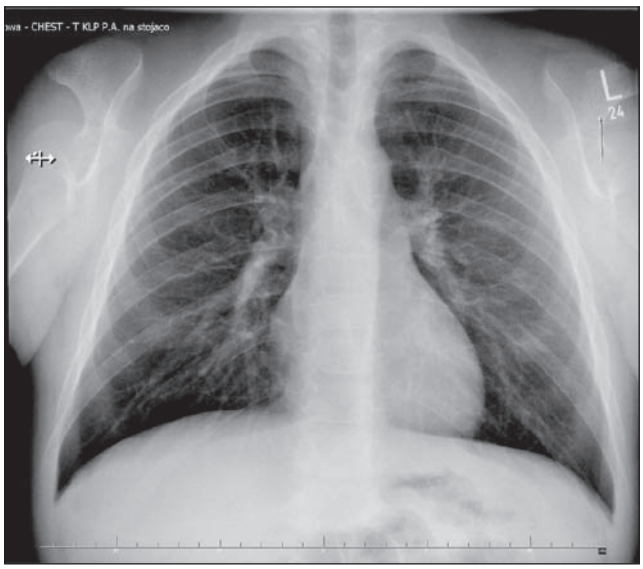

Fig. 3. Chest X-ray confirming the lack of clavicles.
Ryc. 3. Rtg klatki piersiowej potwierdzające brak obojczyków.

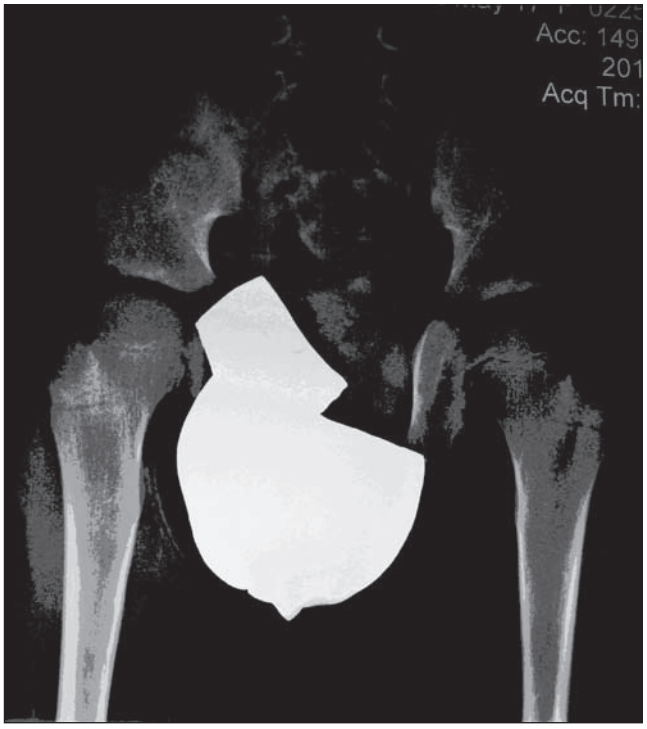

Fig. 4. Pelvic girdle X-ray which shows the acetabular hypoplasia.
Ryc. 4. Zdjęcie obręczy biodrowej, na którym widoczna jest hypoplazja panewek stawów biodrowych.

rib deformation, as illustrated in figure 3. The X-ray of the pelvic girdle made in early childhood showed the acetabular hypoplasia (fig. 4).

The pantomogram and conical tomography showed the presence of retained supernumerary teeth: 13' ,23' ,24' ,33' ,34' ,35' ,43' ,44' ,45' (fig. 5).

The boy was qualified for surgery under general anesthesia. During the operation, which was performed in a typical manner deciduous teeth 55, 65, 74, 75, 84, 85 were removed, then alveoli were mechanically expanded and additional retained teeth 13', 23', 24 ', 33', 34 ', 35' , 43 ', 44', 45 ' were surgically removed (fig. 6 and fig. 7). Lesions corresponding to dentigerous cysts around 13, 24, 35, 44, 45 were enucleated. The postoperative wound was closed with simple interrupted suture. On the second day after the surgery, the boy was discharged and referred for outpatient treatment.

The sutures were removed on the 10th day. The wounds healed properly, intraoral examination revealed eruption of retained teeth 14 and 24. The boy was referred to an orthodontist again.

DISCUSSION

Practice has proved that cleido-cranial dysplasia is a condition which requires highly specialized cooperation of many doctors: a pediatrician, a geneticist, an orthopedic, orthodontic, and maxillofacial surgeon and a prosthetic surgeon. Clinical observations of other authors show

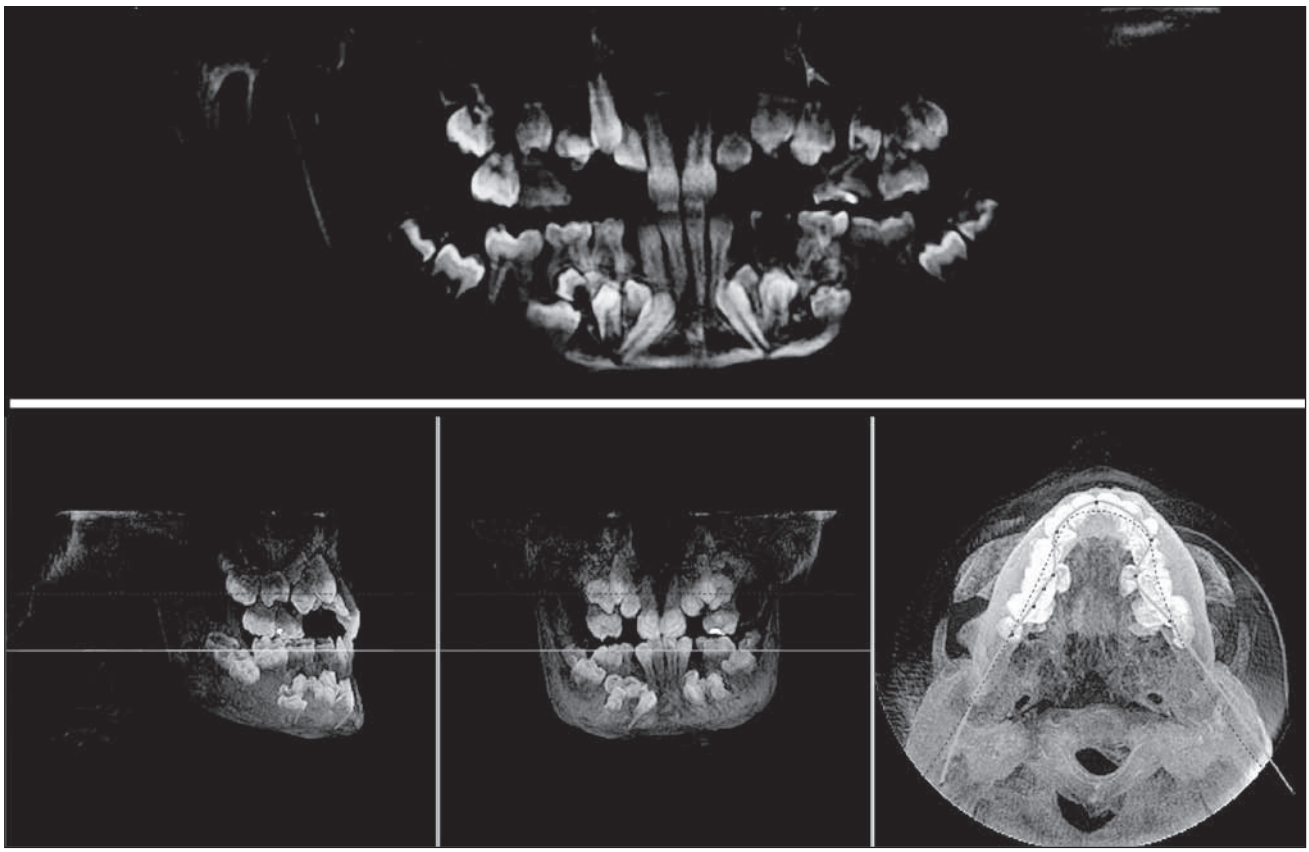

Fig. 5. The Conic CT scan showing the location of supernumerary teeth.
Ryc. 5. Skan tomografii stożkowej ujawniający położenia zębów nadliczbowych.

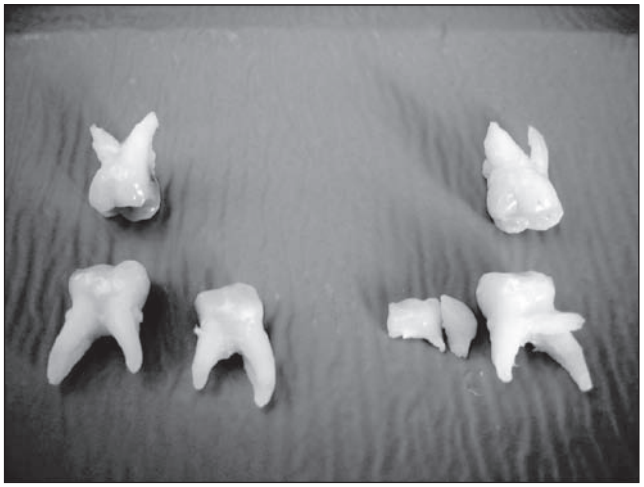

Fig. 6. The picture of removed persistent deciduous teeth.
Ryc. 6. Zdjęcie usuniętych przetrwałych zębów mlecznych.

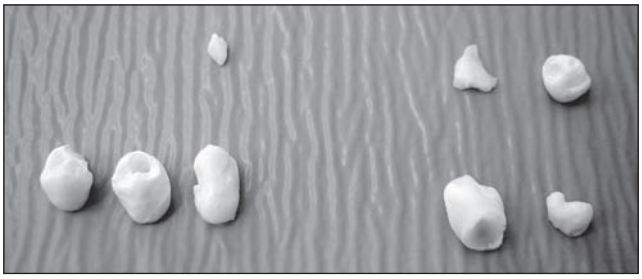

Fig. 7. The picture of removed impacted, additional teeth with characteristic incorrect shape and built.
Ryc. 7. Zdjęcie usuniętych zatrzymanych zębów dodatkowych o charakterystycznych nieprawidłowych kształtach i budowie.

that this disease is also accompanied by anomalies in the masticatory system. The presence of impacted teeth is characteristic of this disease. Koszowski et al reported a case of a CCD patient in whom a total of 30 supernumerary teeth located in the upper and lower jaws were removed [8]. One of the reasons for the irregularities regarding the eruption of permanent teeth results in the lack of root resorption of deciduous teeth and numerous supernumerary teeth, which pose mechanical obstacles. Removing of persistent deciduous and retained supernumerary teeth and exposing right impacted permanent teeth must be considered individually for each patient, while paying attention to the degree of root formation, which should reach 1/3 of the proper root length [7, 10]. Adopting early multidisciplinary treatment of patients diagnosed with CCD ensures good results of treatment, improves the patient's quality of life, self-esteem and the perception of the environment. In the case described, the lack of teeth led to the boy often being ridiculed by peers. The authors believe that early initiation of the treatment is conducive to maintaining the normal function of the masticatory system and peer acceptance.

REFERENCES

1. Gulatis S, Kabra M. Cleidocranial dysplasia. J Posgrad Med. 2001;47:204-205.
2. Garg RK, Agrawal P. Clinical spectrum of cleidocranial dysplasia. A case report. Cases J. 2008;8:377-379.
3. Bhargava P, Khan S, Sharma K, Bhargava S. Cleidocranial dysplasia with autosomal dominant inheritance pattern. Ann Med Health Sci Res. 2014;supp 2: 152-154.
4. Mudlos S. Cleidocranial dysplasia: clinical and molecular genetics. J Med Genet. 1999;36:177-182.
5. Karaguzel G et all; Cleidocranial Dysplasia: A Case Report; J Clin Pediatr Endocrinol. 2010;3:134-136.
6. Mohan RP. Cleidocranical dysplasia clinical-radiological ilustration of rare case. J Oral Si. 2010;57:161-166.
7. Shaikh R, Shusterman S. Delayed dental maturation in cleidocranial dysplasia; J Dent Child. 1998;65:325-329.
8. Koszowski R, Myrda J, Pająk J. Nieprawidłowości szczękowo-zgryzowo- zębowe w przebiegu dysplazji obojczykowo-czaszkowej – opis przypadku. Dent Med Probl. 2004;41:811-816.
9. Rahnama M, Czajkowski L, Świątkowski W, Jachewicz T, Kiełbowicz D. Dysplazja obojczykowo-czaszkowa z mnogim zatrzymaniem zębów. Magaz Stomatol. 2012;7/8:70-72.
10. Nayer G, Ortakoglu K, Senciman M. Multiple impacted teeth. Repoprt of 3 cases. Eur J Dent. 2008;2:73-78.
11. Pospieszyńska M. Ortodontyczne leczenie chorych z nieprawidłowościami uzębienia w zespole obojczykowo-czaszkowym. Dent Forum. 2006;24:87-90.

Author's contributions/Wkład Autorów
According to the order of the Authorship/Według kolejności

Conflicts of interest/Konflikt interesu
The Authors declare no conflict of interest.
Autorzy pracy nie zgłaszają konfliktu interesów.

Received/Nadesłano: 23.06.2015 r.
Accepted/Zaakceptowano: 13.10.2015 r.

Published online/Dostępne online

Address for correspondence:
Bogumił Lewandowski
Clinical Department of Maxillo-Facial Surgery,
Provincial Specialist Hospital in Rzeszów
ul. Chopina 2, 35-005 Rzeszów
Phone 17 866 62 60, 605-547-070
e-mail: boglewandowski@wp.pl