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RARE FACIAL CLEFTS

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In 1976 Dr. Paul Tessier described numeric classification for rare craniofacial clefts. He first emphasized that a fissure of the soft tissue corresponds, as a general rule, to a cleft of the bony structure. The classification, easy to understand, became widely accepted because the recording of the malformations was simple and facilitated communication between observers.

The aim of this study was to present our own experience with treatment of patients with rare facial clefts.

Material and methods. Our Department has 11 patients with rare craniofacial clefts under its care. This group includes 8 boys and 3 girls. The patients aged from 2 months to 18 years at the time of the first consultation.

Results. In two patients the cleft was median, in seven patients it was one-sided and in two – bilateral. The most common type of cleft was number 6, and the rarest were 2, 3, and 7. All patients underwent surgical treatment.

Conclusions. Atypical facial clefts are rare congenital anomalies, however because of functional and aesthetic disturbances they constitute a serious medical and therapeutic problem. Facial clefts are characterized by variable clinical presentation and require individualized treatment plan.

Key words: facial cleft, classification, treatment

In 1976 Dr. Paul Tessier described numeric classification for rare craniofacial clefts (1). Depending on the location of a cleft in relation to the median facial line, he ascribed numbers from 0 to 14 to the defects (2). In his classification he also included eye socket as an area that divided the skull into facial and cerebral skull. Clefts number 0 to 6 are craniofacial defects, while clefts number 8 to 14 are cerebrocranial defects (fig. 1) (3). The cleft number 7 is the most lateral craniofacial cleft. When craniofacial and cerebrocranial malformations coexist, he proposed to extend cleft number 0 to 14, and number 1 to 13, etc. (3). The group of craniofacial defects may also include clefts of the lower lip and maxilla and is collectively termed as cleft number 30 (fig. 1) (4). The author found that a soft tissue cleft corresponds to a similar pathology in the bony structures of the face (1). The classification proposed by Tessier is simple, logical and has been widely accepted due to precise description of a defect and facilitation of comparison of various defects between centers (1).

An estimated incidence of rare craniofacial clefts is 1.43 to 4.85 per 100,000 live births (5). A cleft number 3 is the most common, while malformations number 4 and 5 are the least common (3, 6, 7). Treatment of children with rare craniofacial clefts is very individualized and presents a significant challenge for the medical teams (8, 9). The multispecialist and multistage therapy of a child with the craniofacial cleft involves cooperation of a neonatologist, podiatrist, plastic surgeon and maxillofacial surgeon (9, 10). The treatment course depends on the type of the defect and variability and intensity of these defects is exceptionally high. Adequate imaging diagnostic workup, presenting full spectrum of the disorder, is an important issue. Imaging studies, in

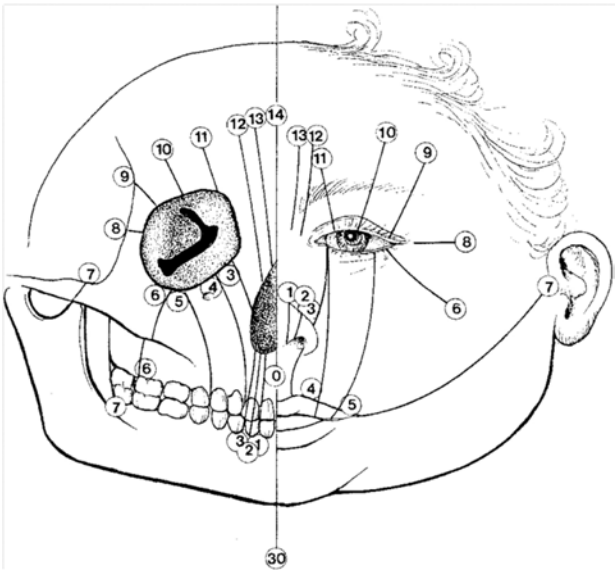

Fig. 1. Classification of the craniofacial clefts according to Tessier; line of the bone tissue cleft is shown on the left side and line of the soft tissue cleft is shown on the right side (3)

particular 3D computed tomography, are very important here since commonly they enable proper diagnosis to be made (9, 11, 12). Cerebrocranial and soft tissue procedures can be performed in the infancy (from 3 to 12 months of life). Reconstruction of the middle part of the face and bone transplantations can be performed in older children (between 6 and 9 years of life). If orthognatic procedures are required, they can be performed when maturity of the craniofacial skeleton is reached (over 14 years of life) (3).

The aim of this study was to present our own experience with treatment of patients with rare facial clefts.

MATERIAL AND METHODS

The Department and Outpatient Clinic of Plastic, Reconstructive and Aesthetic Surgery has 11 patients with rare craniofacial clefts under its care. All patients underwent clinical examination and were photographed. A database of patients with rare facial clefts was created based on medical records, outpatient records and photographs. The collected data included: age, sex, type of the defect, cleft number and method of surgical treatment.

The study group included 8 boys and 3 girls. At the time of their presentation to the Outpatient Clinic, the patients aged from 2 months to 18 years (an average of 2 years and 5 months). The majority of patients continue their care at the Outpatient Clinic until the age of 18 years or more.

Based on medical records of patients with craniofacial clefts and created database, a retrospective analysis of our patients was performed.

No statistical analysis was conducted due to descriptive nature of the study.

RESULTS

Table 1 presents the obtained results. Two patients were diagnosed with a median facial cleft (number 0 according to Tessier), involving the upper lip/small notch of the vermilion and palpable lack of continuity of the orbicularis oris muscle (patient 1) and the upper lip and the maxilla (patient 2). Furthermore, a concealed submucosal palatal cleft was found in the first patient. Both patients underwent plasty of the upper lip in their first year of life; the first patient underwent surgical treatment exactly at the age of 12 months since his patients visited the Outpatient Clinic only when their child was 11 months old. The second patient underwent treatment at the age of 18 months due to wide cleft of the upper lip.

The cleft number 2 according to Tessier was diagnosed in one girl. The defect occurred on the left side, while typical cleft of the lip, alveolar process and the palate was found on the right side. The first stage of surgical treatment, performed at the age of 6 months, involved reconstruction of the left lower lip /larger cleft width/. The second procedure, performed at the age of 1 year, was plasty of the right lip. Palatoplasty was another stage of treatment (at the age of 2 years). Transplantation of a facial bone to the alveolar process of the maxilla was performed at the age of 12 years due to orthodontic indications.

One boy was diagnosed with the cleft number 3 according to Tessier; this defect was found on the left side. Furthermore, typical cleft of the lip, alveolar process and the palate on the right side and drooping of the superior lid of the right eye was diagnosed (fig. 2). The surgical treatment was started with reconstruction of the lip on the left side /6 months/, subsequently on the right side (1 year and 5

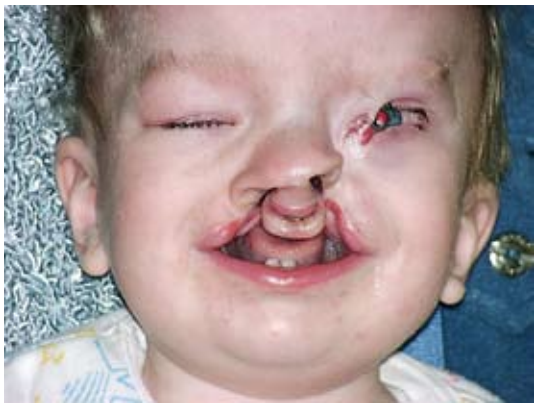

Fig. 2. A boy with left-sided cleft number 3 according to Tessier and typical right-sided cleft of the lip, alveolar process and palate

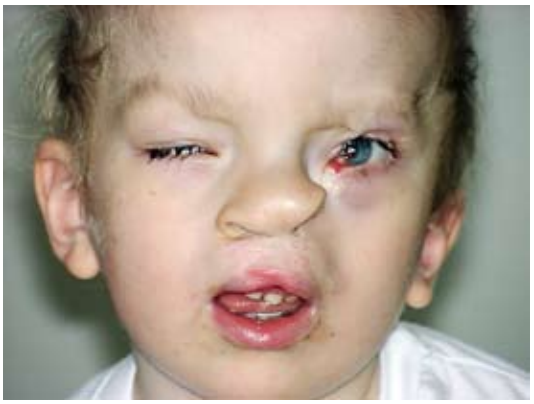

Fig. 3. The same boy after first stages of surgical treatment (bilateral plasty of the upper lip, plasty of the lids of the left eye and palate)

months). At the age of 2 years the corrective plasty of lids of the left eye was done and half year later palatoplasty was performed (fig. 3). At next stages the patient will be qualified to the plasty of the drooping superior lid of the right eye and nose surgery.

Two patients were diagnosed with the cleft number 4 according to Tessier. This defect involved left side in one boy and right side in another one. One of these boys underwent surgical treatment at another center and visited the Outpatient Clinic at the age of 18 years to undergo qualification to the corrective rhinoplasty. The second boy underwent lip plasty on the right side and two years later – a corrective plasty of the superior lid of the right eye.

One boy was diagnosed with bilateral asymmetric oblique facial cleft: number 4 on the right side and number 6 on the left side. Due to his remote residence he underwent surgical treatment at another center.

The next three patients were diagnosed with facial cleft number 6 according to Tessier. Two girls had right sided cleft, while the boy had a bilateral cleft. Furthermore, the boy was diagnosed with a typical cleft of the lip and the alveolar process on the left side (fig. 4). One of the girls underwent her first surgical correction of the oblique facial cleft at another center, while the other girl underwent surgical treatment at our Department at the age of 8 months. Another stages of treatment were similar in both patients (tab. 1). The boy underwent lip reconstruction on the left side at the age of one year and two years later he underwent surgical correction of the oblique cleft of the face. At the age of 8 years he underwent a corrective plasty of the deformed margin of the lower lid of the right eye, and at the age of 12 years – bone transplantation to the alveolar process of the maxilla on the left side (fig. 5A). Due to marked disorder of the maxillary teeth, the patient was referred to an orthopedic treatment in an expedited setting (fig. 5B).

The last of our patients was diagnosed with facial cleft number 7 according to Tessier. The boy underwent surgical treatment at the age of 8 months involving plasty of the lateral facial cleft (tab. 1).

All patients were consulted and/or treated by a maxillofacial surgeon.

DISCUSSION

The clinical presentation of rare clefts is very polymorphic (3). The Tessier's classifica-

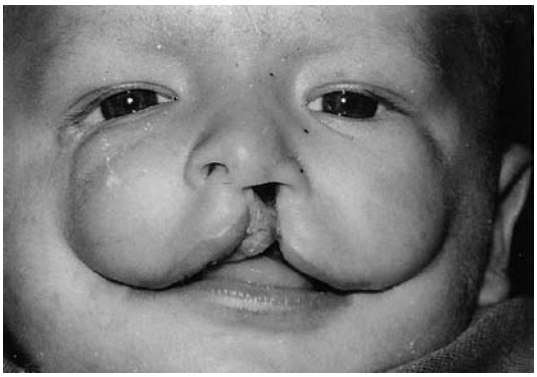

Fig. 4. A boy with bilateral oblique cleft number 6 according to Tessier and coexisting typical cleft of the lip and alveolar process on the left side

Table 1. List of patients with rare facial clefts treated at our Department

No.	Sex	Age	Cleft type	Cleft number	Side	Procedures	Comments
1	M	11 months	middle	0	median	1st year – lip plasty	concealed median lip and palate cleft
2	M	2 months	middle	0	median	8 months – lip plasty	
3	K	6 months	middle	2	l	6 months – lip plasty on the left side, 1st year – lip plasty on the right side, 2 years – palate plasty, 12 years – bone transplantation to the alveolar process	typical cleft of the lip, alveolar process and palate on the right side
4	M	10 months	middle	3	l	11 months lip plasty on the left side, 1 year and 5 months lip plasty on the right side, 2 years – plasty of the lids of the left eye, 2 years and 6 months – palatoplasty	typical cleft of the lip, alveolar process and palate on the right side
5	M	18 years	oblique	4	l	underwent surgical treatment at another center; visited our department to undergo rhinoplasty	
6	M	11 months	oblique	4	r	1st year – lip plasty; 3 years and 6 months – plasty of the lower lid of the right eye	
7	M	2 months	oblique	4 – r 6 – l	bilateral	underwent surgical treatment at another center	palate cleft
8	K	4 years	oblique	6	r	first surgical procedure at another center, 4 years – plasty of the right cheek scar, 16 years – corrective plasty of the deformed margin of the lower lid of the right eye	
9	K	5 months	oblique	6	r	8 months – plasty of the oblique facial cleft, 4 years – plasty of the right cheek scar, 14 years – corrective plasty of the deformed margin of the lower lid of the right eye	
10	M	4 months	oblique	6	bilateral	1st year – lip plasty, 3 years – bilateral plasty of the oblique palate cleft, 8 years – corrective plasty of the deformed margin of the lower lid of the right eye, 12 years – bone transplantation to the alveolar process	typical cleft of the lip and alveolar process on the left side
11	M	6 months	lateral	7	r	8 months – plasty of the lateral facial cleft	

tion is useful and facilitates uniform nomenclature. However, one must bear in mind that there are multiple cleft combinations and that other face defects may coexist. Our material included only craniofacial defects and malformations did not extend to cerebrocranial clefts. Thus clefts number 0 to 7 according to Tessier were diagnosed in our group (tab. 1).

There are reports available that shorten the Tessier's classification of rare facial clefts to three groups: middle clefts (including median clefts) involving numbers 0-3; oblique (4-6) and lateral clefts (otherwise referred to as transverse ones, number 7) (9, 10). In our group the middle facial cleft was found in four patients, oblique – in six and lateral in one subject.

When we compare the same types of clefts among patients, we often found various degrees of the defect. Such defects may be uni- or bilateral and in patients with bilateral defects, one type of defect may be present on one side, while another one on the other side (3). In our material there were two midline clefts, three left sided and four right sided as well as two bilateral clefts.

There is no consensus in the literature whether rare facial clefts are more common in women than in men. Facial clefts were more common in women in the material presented by the Dutch authors (9, 10). However, the defects were more common in male subjects in the group investigated by Alonso et al. and

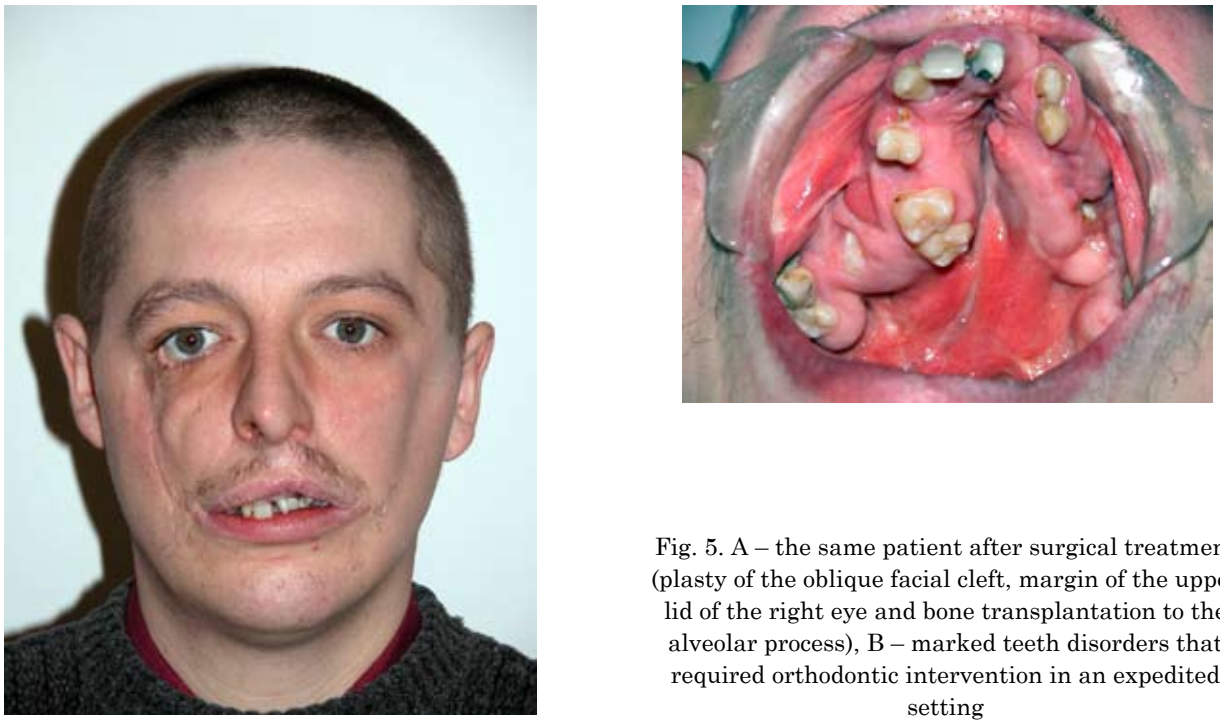

Fig. 5. A – the same patient after surgical treatment (plasty of the oblique facial cleft, margin of the upper lid of the right eye and bone transplantation to the alveolar process), B – marked teeth disorders that required orthodontic intervention in an expedited setting

Allam et al. (13, 14). Similar observations were made in our material where men constituted majority of patients (8 subjects).

Bodin et al. report that the incidence of rare craniofacial clefts is 1.43 to 4.85 per 100,000 live births (5). An average incidence of rare facial clefts, based on our own data, was 4.95 per 100,000 live births. Slightly higher incidence than that reported by Bodin et al. may result from generally higher incidence of clefts in Poland versus in countries of the Western Europe and the USA. However, the above mentioned calculations indicate that these malformations are rare.

Gawrych et al. emphasize possibility of coexistence of a rare facial cleft with a typical cleft of the face, alveolar process and/or palate (15). Lip and/or palate cleft was found to coexist in five patients from the study group (tab. 1). We must emphasize that the palate cleft was submucosal in one patient. This indicates that a detailed physical examination of any young patients with a congenital craniofacial malformation is mandatory.

Treatment of patients with rare facial clefts is very individualized, though some authors tried to create some algorithms of management (9, 10). Surgical treatment of patients with coexisting typical lip and/or palate clefts is the same as in the typical treatment of this defect (16). We must emphasize that in bilateral clefts the surgical treatment should be started at the side where the cleft fissure is wider, which will reduce the tissue tension during the surgical treatment of the opposite side. Soft tissue defects should be corrected in the first year of life of a child (3, 9). Corrections of scars or deformations, in particular those located in the lids, should be performed around the age of 4 years (9). Surgical procedures involving the skeleton (maxilla, nose) are performed in older children (>16 years) (3, 9). Similar management was used in patients from our group and particular attention was put to specificity of particular procedures depending on the defect type and psychomotor development of a young patient.

CONCLUSIONS

1. Atypical facial clefts are rare congenital anomalies, however because of functional and aesthetic disturbances they constitute a serious medical and therapeutic problem.
2. Facial clefts are characterized by variable clinical presentation and require individualized treatment plan.

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