Impact of abnormal construction of the facial surface on speech functions in children with kraniosinostosis - preliminary reports

(Wpływ nieprawidłowej budowy twarzoczaszki na funkcje mowy u dzieci z kraniosynostozami – doniesienia wstępne)

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Abstract – Introduction. Craniosynostosis is congenial malformation caused by premature fusion of one or more cranial sutures leading to abnormal shape of the skull. Because of that non-syndromic craniosynostoses can result in speech disorders
The aim of the study. The aim of the study presented in this study was to assess the incidence of anatomical defects and functional disorders of the orophylax area and speech disorders in children with craniosophthalmitis and frontal, as well as to compare the incidence of these abnormalities in children who underwent surgery to reconstruct the skull with a group of children who did not the surgery was carried out.
Materials and Methods. The authors have performed a neurologopaedic research on a group of 60 children between the ages of 5 and 12 with sagittal and metopic craniosynostoses. 44 of them underwent a reconstruction of the skull and 16 were not operated.
Results. Many children had incorrect structure and functions of speech organs and also speech disorders. In those cases cause of speech disorders is very complex.
Conclusions. Additional research concerning speech and language functions in larger group of school age children with craniosynostoses is needed.

Key words **-** craniosynostosis, speech disorders, articulation.

Streszczenie – Wstęp. Kraniosynostoza jest wadą wrodzoną polegającą na przedwczesnym zarośnięciu jednego, dwóch lub większej liczby szwów czaszkowych, prowadzącą do nieprawidłowej budowy zarówno mózgo- jak i twarzoczaszki. Z tego powodu u dzieci z izolowanymi postaciami kraniosynostoz, może dojść do wystąpienia rozmaitych zaburzeń w zakresie funkcji mowy.
Cel badań. Celem przedstawionych badań była ocena częstości występowania wad anatomicznych i zaburzeń funkcjonalnych obszaru orofacjalnego oraz zaburzeń mowy u dzieci z kraniosynostozą strzałkową oraz czołową, a także porównanie częstości występowania tych nieprawidłowości u dzieci, które przebyły operację rekonstrukcji czaszki z grupą dzieci, u których nie przeprowadzono operacji.
Materiał i metody. Autorzy przeprowadzili badanie neurologopedyczne w grupie 60 dzieci w wieku od 5 do 12 roku życia leczonych z powodu kraniosynostozy strzałkowej oraz czołowej. 44 dzieci przeszło zabieg rekonstrukcji czaszki, a 16 nie było leczonych operacyjnie.
Wyniki. U większości badanych stwierdzono występowanie nieprawidłowości w zakresie budowy i pracy narządów artykulacyjnych oraz zaburzenia mowy, których przyczyna jest bardzo złożona.
Wnioski. Konieczne są dalsze badania dotyczące funkcji mowy w większej grupie dzieci z kraniosynostozami w wieku szkolnym.

Słowa kluczowe – kraniosynostoza, zaburzenia mowy, artykulacja.

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I. INTRODUCTION

The human skull is made up of 22 bones joined by stitches. It consists of two interconnected and interacting parts: the skull, which consists of its upper part - called the vault and the lower part - called the base and the facial skeleton [1] . With flexible seams neonatal cranial bones of the skull can grow properly allowing the brain to normal growth. Craniosynostosis is a congenital defect , which consists in prematurely growing one, two or more cranial sutures [2] . Craniosynostoses may arise against genetic syndromes (eg Crouzon, Apert, Pfeiffer, Saethre-Chotzen, Muenke and many others) - they are then called syndromic craniosynostoses or may not be associated with genetic syndromes - a syndrome they are then isolated (non- syndromic craniosynostoses) [3] . The type of craniosynostosis, and therefore its name, depends on which of the cranial sutures has prematurely become overgrown. We therefore distinguish craniosynostosis by the fibular, frontal, coronary, carbonatic or complex in if more seams are overgrown [4]. Premature occlusion of even one cranial suture leads to abnormal construction of the whole skull, which in turn disturbs the growth and functioning of structures inside, such as the brain, organs of speech, sight and hearing, and skeletal muscles of the head [5]. The influence of isolated craniosynostoses on the child's psychomotor development has been the topic of discussion for many years including the development of speech functions [6, 7, 8] .

The aim of the study presented in this study was to assess the incidence of anatomical defects and functional disorders of the orophylax area and speech disorders in children with craniosophthalmitis and frontal, as well as to compare the incidence of these abnormalities in children who underwent surgery to reconstruct the skull with a group of children who did not the surgery was carried out.

II. MATERIALS AND METHODS

The study group consisted of 60 children treated for coronary amorphisms . The group consisted of 40 children with craniosophthalis (SCP) and 20 with frontal (TRI) . 44 children (30 from SCP and 14 from TRI) in early childhood (between 6 and 15 m.) a cranial reconstruction surgery at the Department of Neurosurgery of the Upper Silesian Children's Health Center or at the Centrum Wad Twarzoczaszki of the Provincial Specialist Children's Hospital in Olsztyn. 16 children (10 with SCP and 6 with TRI) were not treated surgically. Among the surveyed children were 52 boys and 8 girls aged 5 to 12 years, and their average age was 6.48 years (SD: 1.52 r.). All children were diagnosed neurologopedically at the Center for the Treatment of CNS Disorders and Development of Children "Kangaroo".

A detailed neurologopedic diagnostics was performed including an assessment of the construction and motor articulatory organs. The mobility, resting position and tension of the tongue muscles as well as the ability to orientate were examined, as well as the shortening of the sublingual frenulum. The muscles of the cheeks, the orbicularis of the mouth, the hard and soft palate , the condition of the teeth, the position of the mandible and occlusion conditions were examined . The method of breathing, digestive functions such as biting, chewing and mastication, as well as the swallowing pattern were assessed. We examined the feeling in the mouth, oral and kinaesthesia praxis. The level of development of communication and linguistic functions at the semantic, syntactic and inflectional levels were also assessed: the level of comprehension, the vocabulary and the ability to adequately use it in the communication context, dialogue and narrative speech in terms of content and grammar were assessed. The way of articulation of the sounds of a series of skimming was also appreciated: [š], [ž], [č], [ʒ] , hissing : [s], [z], [c], [ʒ] , silent : [ś], [ź], [ć], [ʒ] [and sounds [t], [d], [n], [l] and [r].

Type of craniosynostoses has been confirmed by neuroimaging clinical studies using computed tomography (CT) imaging and three-dimensional background by computer tomograph (3D-CT).

Statistical methods

Statistical analysis was developed using the IBM SPSS Statistics 25 package. Significance of differences between independent groups were estimated using Student's t-test. Significance was assumed at the level of $p < 0.05$.

III. RESULTS

Tables 1-3 present the percentage of children with craniosophthalmic syndrome (SCP) and frontal (TRI) who

had irregularities in the construction or functions within the speech organs.

Table1. Results of the neurologopedic examination

Irregularities in the field	SCP n = 40	TRI n = 20
Hard palate	**100%**	**100%**
Soft palate	32.50%	30.00%
Occlusion	**92.50%**	**90.00%**
Tension mm of cheeks	25.00%	25.00%
Tension mm of the tongue	**42.50%**	30.00%
Tongue mobility	25.00%	30.00%
Rest position of the tongue	**67.50%**	**70.00%**
Tongue verticalization	**50.00%**	40.00%
Sublingual frenulum	37.50%	35.50%

All examined children had normal physical hearing. In 31.66% of subjects, an abnormal oral or mixed airway was found. None of the surveyed children in both groups, were not overt cleft of hard or soft palate, or submucosal cleft, but all patients were found incorrect structure of the hard palate. Most frequently, high arched gothic palates and an asymmetric structure of the hard palate were found. Most children also occurred malocclusion, among which were the most common malocclusion class II by. Angle and cross bites , while open bites and bites in the third grade of Angle were found only in a few cases. Most of these children were during or prior to the beginning of orthodontic treatment. Many patients had narrow jaws and mandible, and in their front sections crowding the incisors. The most frequent diagnoses were hypotonia and asymmetry of muscle tone in the tongue and cheeks, only a few patients showed increased muscle tone of the masseter and temporal muscles, as well as hyperactive muscle muscles.

In most cases, an incorrect resting position of the tongue was stated - the tongue was most often found at the bottom of the mouth , and on the lower incisors, it was often arranged in dysmetry. Some patients underwent undercutting of the sublingual frenulum, and the majority of whom were found to have abnormalities during the current study had a frenulum shortened to a slight or insignificant to medium degree.

Table 2 Results of neurologopedic examination

Irregularities in the field	SCP n = 40	TRI n = 20
Swallowing pattern	**72.50%**	**75.00%**
biting off	7.5%;	10.0%;
chew	25.0%;	25.0%;
chewing	30.0%	30.0%
Compensatory movements of the mandible	30.0%;	25.0%;
Compensatory lip movements	7.5%	15.0%
Sensory in the oral cavity	22.50%	35.00%
kinesthesia	17.50%	5.00%
praxis	17.50%	5.00%

Some of the subjects had disturbances in the scope of food functions, in which the swallowing pattern was most often disturbed. Most often infantile, additive, dorsal and lateral addional swallowing pattern were found.

Table 3. Results of neurologopedic examination

Irregularities in the field	SCP n = 40	TRI n = 20
articulation	**70.00%**	**80.00%**
Articulation of a series of hissing sounds	**45.00%**	**45.00%**
Articulation of a series of noise sounds	**45.00%**	35.00%
Articulation of the sounds of the silence series	**42.50%**	30.00%
Articulation of sounds [t], [d], [n]	**60.00%**	**75.00%**
Articulation of the sound [l]	**50.00%**	**60.00%**
Voice articulation [r]	**50.00%**	**60.00%**

In the majority of patients treated for sagittal craniosynostoses and coronal speech defects were observed under straight dyslalii multiple form, in some cases of complex multiple dyslalia. Most often, defective articulation of sounds [t], [d], [n] and [l] and [r] were found, and the articulation method strongly correlated with the swallowing pattern (Spearman rho 0.830, p = 0.000). Quite often, during the articulation, the occurrence of compensatory movements of the mandible were observed.

In 32.5% of children with craniosophthalmitis and in 20% of children with craniosphthalosis, dysfunction of linguistic functions was found at the inflectional, semantic and syntactic level. It was found to have poor vocabulary inward and outward, and structure of the expression characterized by excessive agrammatisms concerning both the use of unsuitable inflectional endings, as well as the abnormal pattern of words in sentences. 88% of the children examined participated in the past or participated in speech therapy.

There were no statistically significant differences in the incidence of functional disorders and speech disorders in children with sagittal and frontal craniosophthalis.

Among children with frontal craniosynostosis no statistically significant differences were found in the range of abnormalities between children after cranial reconstruction surgery and non-operated children.

There were statistically significant differences between the results obtained in the group of children with craniosophthalis. Children who underwent a reconstruction of the skull in early childhood had significantly better language mobility ($p = 0.021$) , and also significantly less frequent praxis disorders ($p = 0.030$) and kinesthesia ($p = 0.031$) within the articulatory organs than in case of children who were not operated on.

IV. DISCUSSION

Speech belongs to the most complex cognitive functions, and its development depends on many factors, in particular on the properly developing CNS and also on the proper construction and functioning speech organs. In the present study, numerous irregularities in the construction and operation of the articulatory organs have been found. Similarly, previous studies carried away by the authors in a group of 124 children with insulated craniosynostosis [9] many disturbances were observed in basic oral functions, such as incorrect swallowing pattern (38.4%) and irregularities in the functioning of the speech organs, such as the lack of uprightness (81%) and incorrect resting position of the tongue (79%). In the current studies, an abnormal swallowing pattern was found in 75% of subjects, and the increase in the number of disturbances in this range may be due to the fact that previous studies Rożek and Larysz [9] examined patients aged 5-52 months old, and in many cases presented the swallowing pattern could still be considered normative. However, taking into account the fact that the majority of children from the study group underwent speech therapy, the percentage of children with abnormal swallowing pattern should be smaller, as this is a function which, in the case of speech impediments, as also confirmed by current research, should be in the first place corrected. In the context of the prevalence of disorders in the field of food functions, muscular tone in the oral cavity, motorics of the articulatory organs and malocclusion in the group of children with craniosynostoses , it is not surprising that such a high percentage of speech defects (up to 80%). However, in an earlier one evaluation of articulation carried out in a group of 23 children aged 2.5 years to 4.5 years Rożek and Larysz [9] demonstrated the presence of speech defects in up to 91.3%, but this may indicate a tendency towards improvement in speech also effect of speech therapy.

It seems that pre-existing speech disorders in early childhood, including delayed development of active and passive speech, may result in obstruction of language functions, including narrative speech, writing, reading and learning [8, 10]. In current research, these problems have been revealed in over 30% of the examined children. Also, Kapp-Simon *et al*. [10] in 179 children with isolated craniosynostoses in school age showed that they achieved lower scores in language skills and learning compared to healthy controls.

In the present study, speech disorders have been diagnosed , including articulation defects in the majority of patients with craniosophthalmitis and frontal craniosynostosis. Most researchers also report speech disorder in children with isolated craniosynostoses at various ages. Korpilahti *et al*. [8] diagnosed speech disorders in 51% of 61 children with isolated kerriosinostosis . Shipster *et al*. [11] u 37% of 76 children with craniosunostasis found disorders associated with language development. Becker [12] found speech disorders in 49% of 215 patients treated for isolated craniosynostoses , and Larysz [7] in a group of 174 examined children diagnosed retardation or other active speech disorder in 44.3%. Naran *et al.* [13] diagnosed speech disorders in 56.4% of the 100 examined children with isolated craniosynostoses.

In the light of the results presented here concerning the coexistence of many irregularities in the construction and functioning of the broadly understood speech apparatus in children with isolated forms of craniosynostasis, it seems necessary to cover these children with constant neurologopedic care to observe and support speech development, and if necessary to immediately implement effective neurologopedic therapy.

V. CONCLUSIONS

- Most children with sagittal and frontal craniosynosomiasis have speech disorders.
- The incidence and types of functional and speech disorders in children with osteochondral and frontal ostia are similar.
- In children with craniosophthalis who had undergone cranial reconstruction, kinesthesia and oral praxis were significantly less frequent and greater mobility of the tongue was noted than in children not treated surgically with the same disorder.
- Due to the coexistence of disorders in the basic oral functions and speech defects in children with craniosynostoses, it seems necessary to create a paradigm of diagnostic and therapeutic procedures in this group.
- Further studies are needed in a larger group of children at school age treated for isolated intraperitoneal craniosynostoses.

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