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The importance of physical treatment in children underwent craniosynostosis surgery in the first year of life

Značaj habilitacionog tretmana kod dece operisane od kraniosinostoza u prvoj godini života

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Abstract

Background/Aim. Craniosynostosis is a condition that occurs intrauterine or develops in the infant period, and represents premature fusion of cranial sutures. This fusion of sutures limits the normal cranium development and leads to disorder in the phase of rapid growth and development of the brain. Creation of craniosynostoses is associated with an increased incidence of developmental delay during the breastfeeding period. Craniosynostoses are treated by surgery. The role of a physiatrist is to postoperatively assess psychomotor development and implement habilitation treatment. The aim of this study was to determine distribution of the type of craniosynostoses according to the age and gender of patients, effectiveness of habilitation treatment and to estimate the somatosensory evoked potential in the preoperative and postoperative period in children who underwent craniosynostosis surgery in the first year of life. **Methods.** The study was designed as a retrospective research. The data were collected from medical records of 51 children with craniosynostoses and delay in psychomotor development who underwent surgical intervention. The children included in this study, during follow-up, were involved in the habilitation treatment. **Results.** An early diagnosis and surgical intervention had a favorable effect on the development of motor function in children with craniosynostoses. The importance of stimulation treatment in the postoperative period was also proved for achievement of an adequate degree of motor development in children in relation to age. The results of our study confirmed the results obtained in previously published studies that the children who did not undergo surgery and start with the habilitation treatment immediately after it, had delay in psychomotor development of moderate degree. **Conclusion.** Habilitation treatment significantly reduced the deviations in psychomotor development of children with craniosynostoses if it started immediately after the surgical procedure.

Key words:
craniosynostoses; skull; infant; psychomotor disorders; evoked potentials; somatosensory physical therapy modalities.

Apstrakt

Uvod/Cilj. Kraniosinostoza je stanje koje nastaje intrauterino ili se razvija u odojačkom periodu, a predstavlja prerano srastanje kranijalnih sutura. Ovakvo srastanje sutura ograničava normalan razvoj kranijuma i dovodi do poremećaja u fazi brzog rasta i razvoja mozga. Nastanak kraniosonostoza povezan je sa povećanom incidencijom kašnjenja u razvoju tokom odojačkog perioda. Tretman kraniosinostoza je hirurški. Uloga fizijatra je da u postoperativnom periodu izvrši procenu psihomotornog razvoja i sprovede habilitacioni tretman. Cilj ovog rada je bio da se kod dece operisane od kraniosonostoza u prvoj godini života utvrdi distribuciju tipa kraniosinostoza prema uzrastu i polu deteta, utvrdi efikasnost habilitacionog tretmana i ispitaju somatosenzorni evocirani potencijali u preoperativnom i postoperativnom periodu. **Metode.** Sprovedena je retrospektivna studija, a podaci su prikupljeni iz medicinske dokumentacije 51 deteta kod kojih je postavljena dijagnoza kraniosinostoze i urađena hiruška intervencija, a kod kojih je ustanovljeno kašnjenje u motornom razvoju u odnosu na uzrast i tip kraniosinostoze. Deca obuhvaćena ovim istraživanjem su, tokom praćenja, bila uključena u habilitacioni tretman. **Rezultati.** Pokazano je da rano dijagnostikovanje i hirurška intervencija imaju

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povoljan efekat na razvoj motornih funkcija dece obolele od kraniosinostoze. Takođe je dokazan značaj stimulacionog tretmana u postoperativnom periodu na dostizanje adekvatnog stepena motornog razvoja deteta u odnosu na starosnu dob. Rezultati našeg istraživanja potvrđuju rezultate dobijene u ranije objavljeni studijama, da deca koja nisu operisana i uključena u habilitacioni tretman postoperativno, pokazuju kašnjenje u psihomotornom razvoju umerenog stepena.

Zaključak. Habilitacioni tretman značajno je smanjio odstupanja u psihomotornom razvoju dece sa kraniosinostozom, ukoliko je započet odmah posle hirurške intervencije.

Ključne reči:
sinostoze; lobanja; novorođenče; psihomotorni poremećaji; evocirani potencijali, somatosenzorni; fizikalna terapija.

Introduction

Craniosynostosis is a condition that occurs intrauterine or develops in the infant period. It represents premature fusion of cranial sutures [1]. This fusion of sutures limits the normal cranium development and leads to disorder in the phase of rapid growth and development of the brain [2]. The prevalence of craniosynostosis in the United States is 10–16 per 10,000 live births, in the UK 4–5 per 10,000 live births, while in France is 4.7 per 10,000 live births [3]. In the Republic of Serbia, the official data have not been released yet. The clinical picture of craniosynostosis shows disrupted form of the skull (dyscrania), fontanelles disappear little earlier than it is normally, there is a ridge along the ossified suture. Craniosynostosis can be syndromic (Syndrome Apert and Syndrome Crouson) and nonsyndromic. According to the localization, craniosynostoses can be sagittal, metopic, coronal, lambdoid, and combined. According to the etiology, craniosynostoses can be primary, secondary and syndromic. According to the number of affected sutures, they can be a single (solitary) and multiple [4, 5]. Craniosynostoses are treated with surgery, by variety of approaches (frontoorbital improving or endoscopic) depending on the localization [6]. Creation of craniosynostoses is associated with an increased incidence of developmental delay during the breastfeeding period. Motor skills have proved especially vulnerable to damage during this developmental period [2]. Treatment of craniosynostosis is surgical. The role of a physician is to postoperatively assess psychomotor development and implement habilitation treatment. Stimulation treatment should make a favorable impact on the physical and mental development of the child who underwent craniosynostosis surgery in the first year of life, ie. during the breastfeeding period [7].

The aim of this study was to determine distribution of the type of craniosynostosis according to the age and gender of patients, effectiveness of habilitation treatment and to estimate somatosensory evoked potentials (SEP) in the preoperative and postoperative period in children who underwent craniosynostosis surgery in the first year of life.

Methods

The study was designed as a retrospective research; the data were collected from medical records of 51 children with diagnosed craniosynostosis who underwent surgical intervention at the Department of Neurosurgery of the University Children's Hospital in Belgrade in the period from 2011 to 2013. Children had a delay in gross motor development in relation to the age and type of craniosynostosis. Delay in motor development in children was revealed by examination of physiatrists and neurosurgeons, who found that children had not reached a milestone in motor development for their age according to the Munich Development Scale [8]. This scale covers the monitoring of development segments during the first year of life like crawling, sitting, walking, grasping, perception, speech perception and social behavior. Severity of the clinical picture, ie. degree of central coordination disorder, was determined by the physiatrist in relation to the deviation from postural Vojta reactions [9]. The children included in this study, during follow-up, were involved in the habilitation treatment recommended by physiatrists and neurosurgeons.

Dependent variables monitored in the study were: age of a child, preoperative findings of a physiatrist, preoperative findings of SEP, findings of a physiatrist and SEP three and six months after craniosynostosis surgery, while the independent variable was the type of craniosynostosis. In order to apply statistical and econometric methodology, the data relating to the variables were coded as follows: gender: 1 – boy, 2 – girl; craniosynostosis type: 1 – syndromic, 2 – nonsyndromic; type of craniosynostosis: 1 – sagittal, 2 – lamboid, 3 – coronal, 4 – metopic and 5 – combined; findings of a physiatrist: 1 – without findings, 2 – findings of is normal, 3 – disorder of the central coordination of the lower degree (the deviation of the normal motor development ± one month), 4 – disorder of the central coordination of a moderate level (the deviation of the normal motor development 2–3 months), 5 – disorder of the central coordination of severe degree (deviation from the normal motor development up to 6 months); SEP findings: 1 – no finding, 2 – normal finding, 3 – disorder of the lower degree, 4 – disorder of moderate degree; genetic anomaly: 1 – does not exist, 2 – exists.

Results

Our study tested and followed 51 children with craniosynostosis. Of all the respondents, 34 (66.7%) were boys and 17 (33.3%) were girls. The average age of the children expressed through the arithmetic mean was 169.08 days (50% of children were younger than 154 days, and other half of children was older than 154 days). Distribution of patients according to the type of craniosynostosis and gender is given in Table 1. Based on the data presented in Table 1 only one girl had the type 1 of craniosynostosis (syndromic). All other children, 50 of them, had the type 2 of craniosynostosis (nonsyndromic). Among them, 34 (66.7%) were boys and 17

(33.3%) were girls. Distribution of patients according to the type of craniosynostosis and age is shown in Figure 1. The patients were divided into three groups according to their age. The youngest patients, younger than 126 days, mostly suffered from nonsyndromic craniosynostosis type 1 and type 5 (9 or 17.6%, and 8 or 15.7%, respectively). Patients aged between 127 to 182 days mostly suffered from nonsyndromic craniosynostosis type 5 (6 or 11.8%). The same was in the third age group, over 183 days. Ten of them (19.6%) had nonsyndromic craniosynostosis type 5.

Table 1
Distribution of patients based on the gender and the type of craniosynostosis

Type of craniosynostosis	Patients, n		Total, n (%)
	boy	girl	
Syndromic	0	1	1 (2)
Nonsyndromic	34	16	50 (98)
Total, n (%)	34 (66.6)	17 (33.3%)	51 (100)

Distribution of patients with craniosynostoses according to the preoperative findings of a physiatrist related to the central coordination disorder degree is given in Table 2. It was shown that 17 (33.3%) patients had some type of nonsyndromic craniosynostosis but without preoperative finding of a physiatrist. Finding of a physiatrist for one patient was normal, 8 (15.7%) patients had disorder of the central coordination of the lower degree, 19 (37.3%) patients had disorder of the central coordination of the moderate degree and 6 (11.8%) of the patients had disorder of the central coordination of the severe degree. Totally, 33 (64.7%) of the patients with nonsyndromic craniosynostosis had disturbed central coordination of various degrees.

Distribution of patients with craniosynostosis according to the findings of a physiatrist related to the central coordination disorder degree three months after operation is given in Table 3. It was shown that 8 (15.7%) of the patients did not have finding of a physiatrist three months after surgical treatment (5 of them had combined type of craniosynostosis), 3 (5.9%) of the patients had normal finding of a physiatrist, 14 (27.5%) of the patients had disorder of the central coordination of the lower degree, 21 (41.2%) of the patients had disorder of the central coordination of the moderate degree, and 5 (9.8%) of the patients had disorder of the central coordination of the severe degree; a total of 40 (78.4%) of the children had some degree of the central coordination disorder three months after operation, mostly those with sagittal and combined type of craniosynostosis (14 and 18, respectively).

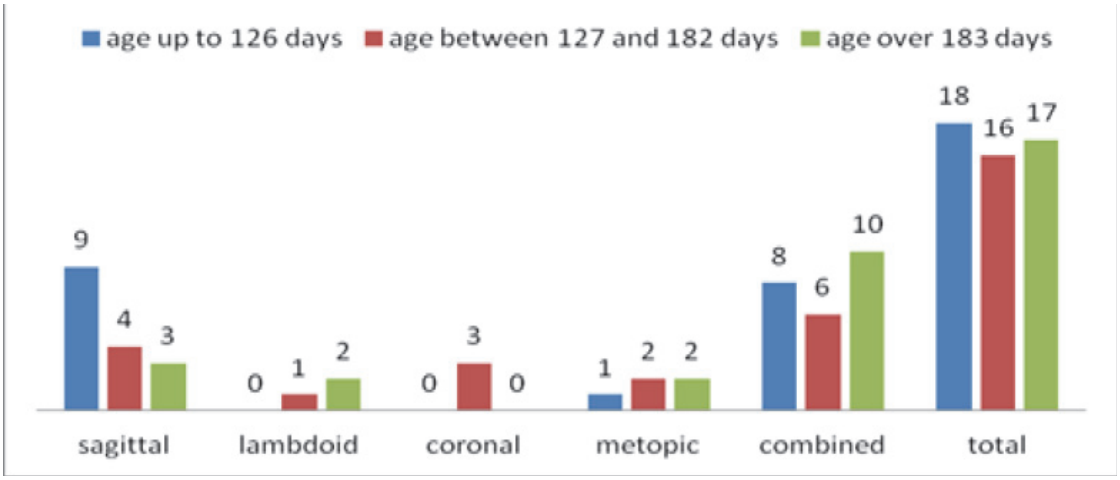

Fig. 1 – Distribution of patients based on the age and the type of craniosynostosis.

Table 2
Distribution of patients based on the type of craniosynostosis according to preoperative findings of a physiatrist

Type of nonsyndromic craniosynostosis	Findings of a physiatrist					Total number
	1	2	3	4	5	
Sagittal	**5**	0	3	**6**	**2**	16
Lambdoid	2	0	0	0	1	3
Coronal	0	0	0	2	1	3
Metopic	2	1	0	1	1	5
Combined	**8**	0	**5**	**10**	1	24
Total number	**17**	**1**	**8**	**19**	**6**	**51**

Note: bolded values represent the highest number in a total sample of patients (n = 51)
Findings of a physiatrist: 1 – without finding, 2 – finding is normal, 3 – disorder of the central coordination of lower degree (deviation of the normal motor development ± one month), 4 – disorder of the central coordination of moderate degree (the deviation of the normal motor development 2–3 months), 5 – disorder of the central coordination of severe degree (deviation from the normal motor development up to 6 months).

Table 3
Distribution of patients based on the type of craniosynostosis according to the findings of a physiatrist three months after surgery

Type of craniosynostosis	Findings* of a physiatrist					Total number
	1	2	3	4	5	
Sagittal	1	1	**6**	**6**	**2**	16
Lambdoid	1	1	0	0	1	3
Coronal	0	0	0	3	0	3
Metopic	1	0	0	3	1	5
Combined	**5**	1	**8**	**9**	**1**	24
Total number	**8**	3	**14**	**21**	**5**	51

Note: bolded values represent the highest number in a total sample of patients (n = 51)
***For explanation see under Table 2.**

Distribution of patients based on the type of craniosynostosis according to the findings of a physiatrist six months after the surgery is given in Table 4. It can be seen that six months after the operation, all the patients had findings of a physiatrist related to the central coordination disorder, mostly the lower and moderate degree (37 patients or 72.5%).

Table 4
Distribution of patients based on the type of craniosynostosis according to the findings of a physiatrist six months after surgery

Type of craniosynostosis	Findings* of a physiatrist			
	2	3	4	Total number
Sagittal	5	4	**7**	16
Lambdoid	1	1	1	3
Coronal	0	1	2	3
Metopic	1	3	1	5
Combined	**8**	**13**	3	**24**
Total number	15	22	14	51

Note: bolded values represent the highest number in a total sample of patients (n = 51)
***For explanation see under Table 2.**

There were no statistically significant differences in distribution of patients according to SEP findings in three different periods of time: preoperatively, three months and six months after the surgical treatment (Table 5).

Discussion

Our study included 51 children, 34 (66.7%) boys and 17 (33.3%) girls, underwent craniosynostosis surgery in the first year of life. This gender distribution of children with craniosynostosis is in accordance with results of the longitudinal studies of American pediatric neurosurgeons, published in 2012 and 2015 [10, 11]. Namely, in those studies two-thirds of children who underwent craniosynostosis surgery were male.

Our results showed that average age of the children was 169.08 days. Previous studies suggest that surgery before the age of 6 months results in the improvement in long-term neurological outcome [12].

Distribution of our patients according to the type of craniosynostosis and gender is consistent with results reported in 2015 that 8% of all the patients affected by craniosynostosis suffer from nonsyndromic craniosynostosis [13]. Most our patients, 16 (31.4%) and 24 (47.1%) had nonsyndromic craniosynostosis type 1 and type 5, respectively.

Published data show that children with detected craniosynostosis usually undergo the surgery at the age of 4 to 16 months [2, 12]. This attitude of American neurosurgeons is different from the attitudes of our neurosurgeons. Our opinion is that the best period form performing the surgical treatment is the age of 0 to 6 months because the first year of life is a time when the habilitation treatment after surgery has the best effect on the of psychomotor development of the child. Craniosynostosis in elderly children cause damage of various cognitive functions (attention, speech, abstract thinking) and their recovery is incomplete after surgery, as evidenced by a number of longitudinal studies [10, 14].

The results after three months of the intervention showed that the number of patients with no findings of a physiatrist reduced from 33.3% to 15.7%. It is necessary that all children underwent the craniosynostosis surgery have physiatrist's examination and habilitation treatment as well. This is the only way that patients with craniosynostosis get the opportunity to achieve maximum functional recovery period [15].

Table 5
Distribution of patients according to sensory evoked potential (SEP) findings, preoperatively, three months and six months after the surgery

SEP findings	Patients, n (%)		
	preoperatively	3 months after surgery	6 months after surgery
No finding	40 (78.4)	39 (76.5)	37 (72.5)
Normal	0 (0)	0 (0)	3 (5.9)
Lower degree disorder	3 (5.9)	6 (11.8)	8 (15.7)
Moderate degree disorder	8 15.7	6 (11.8)	3 (5.9)
Total	51 (100)	51 (100)	51 (100)

Six months after the operation, all the patients had findings of a physiatrist related to the central coordination disorder, mostly the lower and moderate degree.

All the subjects in this study were included in the postoperative habilitation treatment, which led to an evident clinical improvement six months after the intervention. Limitation of our research was the fact that it was not possible to use the scales for evaluation of psychomotor development such as the Psychomotor Development Index as well as the Bayley Scales of Infant Development, which would make physical treatment findings more accurate.

A small number of the SEP findings, regardless of the severity of the disorder and the type of craniosynostosis, show an inadequate diagnosis and monitoring of recovery of patients after the surgery. This result speaks about the need for education of physiatrists about the importance of neurophysiological testing the patients with craniosynostosis.

Our study analyzed the findings of somatosensory evoked potentials. Since the evoked potentials diagnose changes in the conductivity of the afferent fibers and maturation of CNS, the findings that are generated by the SEP represent a reliable parameter of general motor status, and, accordingly, it is expected that children with deviations in the SEP findings are on a continuous rehabilitation. Our research was carried out during the postoperative habilitation. It was shown that the abnormal SEP finding is an important diagnostic tool in assessing the planning and implementation of continuous habilitation. The real significance of neuropsychological testing would be evident if the analysis of all the types of evoked potentials in patients are carried out for a longer follow-up. On this manner an adequate monitoring of the recovery and development of CNS functions would be provided.

The preoperative findings of a physiatrist as well as those 3 months after the surgery did not differ. In both periods, most patients had the moderate degree of the central coordination disorder (19 patients preoperatively and 21 patients 3 months after the surgery). These results suggest that a longer interval of time is required for the physical therapy treatment, regardless of the severity of the clinical findings. Results relating to physical therapy cannot be detected after only three months of the treatment.

Analysis of findings of a physiatrist 6 months after the surgery showed that all the patients had that finding and that was no patient with the central coordination disorder of the severe degree. Besides, this analysis revealed that most of the patients (22 or 43.1%) had the mild impairment of the central coordination, 15 (29.54%) of the patients had normal finding, and 14 (27.4%) of the patients had the central coordination disorder of the moderate degree. These results were much better than those from the preoperative period and the three-months period after the surgery and are consistent with those from the literature [2]. On the other hand, these results show the effectiveness of postoperative habilitation treatment.

Analysis of the SEP findings showed that the most patients had no such a finding in all three period of time when clinical estimation of the patients was performed (78.4%, 76.5% and 72.5% of the patients in the preoperative period, three months after the surgery and six months after the surgery, respectively). Contrary to rhis, the preoperative findings of a physiatrist could not be found in 17 patients whereas 6 months after the surgery all the respondents had such the finding. Due to this we could not perform correlation analysis between clinical and SEP findings in our patients. In order to make the data comparable, it is necessary that a physiatrist at each stage of the treatment refer patients to the neurophysiological testing (preoperatively, three and six months after the surgery).

Regardless of mentioned limitations, our study showed that the early diagnosis and surgical intervention had a favorable effect on the development of motor function in children with craniosynostosis. The importance of habilitation treatment in the postoperative period was also proved for the achievement of an adequate level of motor development in children underwent craniosynostosis surgery in relation to age.

Conclusion

If children with craniosynostosis begin habilitation treatment immediately after the surgical intervention, there is a significant reduction of the psychomotor development deviations.

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