

Onderzoek naar gedrag en hersenfuncties bij kinderen met een craniofaciale aandoening.

No registrations found.

Ethical review	Positive opinion
Status	Pending
Health condition type	-
Study type	Observational non invasive

Summary

ID

NL-OMON21615

Source

Nationaal Trial Register

Health condition

Craniosynostosis is a congenital malformation characterized by premature closure of cranial sutures. The premature closure of the cranial sutures hinders the growth of the skull, brains and face. Craniosynostosis is 1 in 2500 newborns and is for approximately 40% of patients a part of a syndrome such as Apert syndrome, Crouzon / Pfeiffer, Saethre-Chotzen and Muenke. The treatment of syndromic or complex craniosynostosis craniofacial comprises a correction within the first year of life. Depending on the syndrome, multiple corrections of the skull, face hands and feet occur. Besides the appearance, the skull abnormality, hand and foot abnormalities, and brain abnormalities may occur. These brain abnormalities can be congenital, such as abnormalities of the corpus callosum or acquired, such as hydrocephalus. Craniosynostose is een congenitale afwijking gekarakteriseerd door premature sluiting van de schedelnaden. De premature sluiting van de schedelnaden belemmert de groei van de schedel, hersenen en gezicht. Craniosynostose komt bij 1 op de 2500 neonaten voor en is bij ongeveer 40% van de patiënten een deel van een syndroom, zoals het syndroom van Apert, Crouzon/Pfeiffer, Muenke en Saethre-Chotzen. De behandeling van syndromale of complexe craniosynostose bestaat uit een craniofaciale correctie voor het eerste levensjaar. Afhankelijk van het syndroom kunnen meerder correcties aan schedel, gezicht handen en voeten plaatsvinden. Naast de uiterlijke kenmerken, de schedelafwijking, hand- en voetafwijkingen, kunnen afwijkingen in de hersenen voorkomen. Deze hersenafwijkingen kunnen aangeboren, bijvoorbeeld afwijkingen van het corpus callosum of verworven zijn, bijvoorbeeld hydrocephalus.

Sponsors and support

Primary sponsor: Erasmus MC
Rotterdam
the Netherlands

Source(s) of monetary or material Support: Nuts-OHRA
the Netherlands

Intervention

Outcome measures

Primary outcome

Neuropsychological functioning as mentioned in the intervention box. Results on the questionnaires completed by the parents/cargivers.

Secondary outcome

Predictors:

1. Hypoplasia of the corpus callosum, septum pellucidum, hippocampus and cerebral cortex, white matter lesions and brain volume;
2. Ventriculomegaly, chronic tonsillar herniation, increased intracranial pressure, hydrocephalus and impressiones digitatae;
3. Obstructive sleep apnea syndrome (OSAS);
4. Genotype and diagnosis;
5. Age at first craniofacial correction and the type of correction, fronto-orbital advancement versus occipital expansion;
6. Socioeconomic status, child gender, child age.

Study description

Background summary

“The neurocognitive functioning and behaviour of children aged six to twelve years with a syndromic or complex craniosynostosis”.

Craniosynostosis is a congenital malformation characterized by premature closure of cranial sutures. The premature closure of the cranial sutures hinders the growth of the skull, brains and face. Craniosynostosis is 1 in 2500 newborns and is for approximately 40% of patients a part of a syndrome such as Apert syndrome, Crouzon / Pfeiffer, Saethre-Chotzen and Muenke. The treatment of syndromic or complex craniosynostosis craniofacial comprises a correction within the first year of life. Depending on the syndrome, multiple corrections of the skull, face hands and feet occur. Besides the appearance, the skull abnormality, hand and foot abnormalities, and brain abnormalities may occur. These brain abnormalities can be congenital, such as abnormalities of the corpus callosum or acquired, such as hydrocephalus.

The neurocognitive functioning and behaviour of patients with syndromic or complex craniosynostosis remains still unclear. The number of studies of the neurocognitive functioning and behaviour of children with syndromic or complex craniosynostosis is small. Most of these studies exhibit methodological limitations, such as small research using non-validated and standardized instruments are not. Despite methodological limitations, all studies seem to indicate that children with syndromic or complex craniosynostosis an increased risk of learning disabilities and behavioural problems.

The objective of this research project focuses on the cognitive functioning of children with syndromic or complex craniosynostosis, we will investigate the frequency of behavioural problems in these children. Clinical findings presume that the executive functioning, attention and memory are less developed in comparison with the general population, with standardised questionnaires and instruments we will figure out if this assumption is true.

Furthermore we are interested in the medical and child factors, namely brain anomalies, obstructive sleep apnea, genotype, diagnosis and their prediction on the neurocognitive outcome.

The psychological assessment of children takes place in the outpatient department of Child and Adolescent Psychiatry at the Erasmus MC-Sophia Rotterdam the Netherlands. The study is divided into two sessions conducted. All tests are conducted according to a standard. The tests are possible in the same order will be used

The questionnaires are sent to the parents beforehand. The questionnaires are completed and then returned home for research. The TRF is the teacher handed to the parents. This completes the questionnaire and send it in an enclosed return envelope.

Statistical analysis: Data is entered into a database (windows office excel 2007) and using a statistical program worked (SPSS 15.0 for Windows 2000 and GraphPad Prism 4).

The average IQ of the group will be compared with that of the standard group by an independent t-test.

Internalizing, externalizing and total behavior will be compared with the control group using an independent t-test.

The (neuro) cognitive functioning in children with syndromic or complex craniosynostosis will be compared with the control group using an independent t-test.

The correlation between the dependent variables (intelligence, behavior and neurocognitive function) and independent (hypoplasia of the corpus callosum, septum pellucidum, hippocampus, cerebral cortex, white matter lesions and brain volume, ventriculomegaly, chronic tonsillar herniation, increased intracranial pressure, impressiones digitatae, hydrocephalus, OSAS, genotype, diagnosis, age at first craniofacial correction and type of correction) will be analyzed with a Spearman correlation procedure as a dichotomous variable is concerned with a Pearson correlation for the variables that a linear relationship assumption and a ratio or interval measurement scale.

Study objective

Clinically, children with syndromic craniosynostosis seem to have an increased risk of learning disabilities and behavioral problems in comparison with the general population.

Study design

Children undergo the diagnostic procedure only once.

Intervention

1. WISC-III-NL;
2. ANT 2.1;
3. FLANKER;
4. n-BACK;
5. TIME-TEST;
6. SSRT;
7. CBCL;
8. DBD;
9. i-DISC-IV;

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Eligibility criteria

Inclusion criteria

All children receiving treatment at the Cranio-Facial Center ErasmusMC-Sophia with syndromic craniosynostosis (such as Apert syndrome, Crouzon/Pfeiffer syndrome, Muenke syndrome, Saethre-Chotzen syndrome) or a complex craniosynostosis and aged between 6 and 13 years.

Exclusion criteria

1. Children with syndromic craniosynostosis, whose parents master the Dutch language insufficiently to independently complete questionnaires;
2. Children familiar with one another syndromic abnormality.

Study design

Design

Study type:	Observational non invasive
Intervention model:	Parallel
Allocation:	Non controlled trial
Masking:	Open (masking not used)
Control:	N/A , unknown

Recruitment

NL	
Recruitment status:	Pending
Start date (anticipated):	01-01-2008
Enrollment:	85
Type:	Anticipated

Ethics review

Positive opinion	
Date:	27-05-2011
Application type:	First submission

Study registrations

Followed up by the following (possibly more current) registration

No registrations found.

Other (possibly less up-to-date) registrations in this register

No registrations found.

In other registers

Register	ID
NTR-new	NL2779

Register	ID
NTR-old	NTR2919
Other	METC ErasmusMC Rotterdam : 2005-273
ISRCTN	ISRCTN wordt niet meer aangevraagd.

Study results

Summary results

Obstructive sleep apnea-specific quality of life and behavioral problems in children with syndromic craniosynostosis.

Bannink N, Maliepaard M, Raat H, Joosten KF, Mathijssen IM.

J Dev Behav Pediatr. 2011 Apr;32(3):233-8.

Reliability and validity of the obstructive sleep apnea-18 survey in healthy children and children with syndromic craniosynostosis.

Bannink N, Maliepaard M, Raat H, Joosten KF, Mathijssen IM.

J Dev Behav Pediatr. 2011 Jan;32(1):27-33.

Health-related quality of life in children and adolescents with syndromic craniosynostosis.

Bannink N, Maliepaard M, Raat H, Joosten KF, Mathijssen IM.

J Plast Reconstr Aesthet Surg. 2010 Dec;63(12):1972-81. Epub 2010 Mar 10.

Surviving meningococcal septic shock: health consequences and quality of life in children and their parents up to 2 years after pediatric intensive care unit discharge.

Buyse CM, Raat H, Hazelzet JA, Hop WC, Maliepaard M, Joosten KF.

Crit Care Med. 2008 Feb;36(2):596-602.

Long-term health status in childhood survivors of meningococcal septic shock.

Buyse CM, Raat H, Hazelzet JA, Hulst JM, Cransberg K, Hop WC, Vermunt LC, Utens EM, Maliepaard M, Joosten KF.

Arch Pediatr Adolesc Med. 2008 Nov;162(11):1036-41.