# Optimal timing and technique for surgical correction of peri-orbital osseous deformities in congenital craniofacial disorders

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Primary objective:To determine the most optimal technique and timing of surgery for correction of peri-orbital osseous deformities (hypertelorism, vertical orbital dystopia, and/or midface hypoplasia) for each specific condition (1. syndromic...

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| **Ethical review** | Approved WMO |
| **Status** | Will not start |
| **Health condition type** | Congenital and hereditary disorders NEC |
| **Study type** | Observational non invasive |

## Summary

### ID

NL-OMON42580

**Source**
ToetsingOnline

**Brief title**
Outcome following surgical correction of peri-obital deformities

### Condition

- Congenital and hereditary disorders NEC

**Synonym**
congenital facial deformity, syndromic craniosynostosis

**Research involving**
Human

### Sponsors and support

**Primary sponsor:** Mondziekten, Kaak- en Aangezichtschirurgie

**Source(s) of monetary or material Support:** Ministerie van OC&W

## Intervention

**Keyword:** hypertelorism, midface hypoplasia, surgery, vertical dystopia

## Outcome measures

### Primary outcome

Peri-orbital functions

- Visual acuity: the visual acuity (logMAR-chart) and visual field is assessed;

the performed tests are age-dependent.

- Ocular motility: An orthoptist will assess movement with specific attention

for A and V patterns in the nine positions of gaze in a standardized way.

Causes of strabismus will be evaluated.

- Closure and position of eyelids: the presence of lagophtalmus, exorbitism,

enophtalmus, ectropion, entropion, downward slanting, or scleral show, dystopia

of the medial and lateral canthi, and the presence of epicanthus will be

assessed.

- Functioning of the lacrimal system/adnexen.

Peri-orbital esthetic appearance

- Overall appearance: This will be assessed with the Versnel scoring list based

on standardized photographs.

- Hypertelorism: The interdacryon distance will be analyzed on CT or CBCT. In

addition intra-operative measurements of the interdacryon distance in the

patient will be performed before and after correction by the surgeon with a

ruler.

At the level of soft tissue, hypertelorism will be studied on the (3D-) photographs. Intercanthal and interocular distances, and horizontal axis of the eye will be measured.

- Vertical orbital dystopia: Vertical position of the orbits/ orbital floor will be analysed on CT/CBCT (bone) and (3D-)photographs (soft tissue).
- Midface hypoplasia: The advancement of the midface due to surgery or growth, will be studied on cephalometry calculating SNA. On lateral photographs facial divergence will be analyzed drawing a line from the glabella to the upperlip, to the vertex of the soft-tissue chin.
- Additional peri-orbital deformities will be assessed on standardized photographs according to the Versnel score.

Movements of the soft tissue and bone structures will be measured on pre- and postoperative CT/CBCT in millimetres.

Relapse and growth. Relapse will be evaluated in the operated patients. Growth will be analyzed using X-ray and photographs at 0, 4, 9 and 18 years old.

**Secondary outcome**

Postoperative satisfaction of the surgeons will be analysed using a panel. Photographs pre-operatively, 1 year postoperatively and at the age of 18-21 years will be analysed. Surgeons will score surgical outcomes using a VAS. With the VAS hypertelorism, vertical orbital dystopia and midface hypoplasia, the overall appearance, the frontal, orbital, nose and midface region will be judged.

Postoperative satisfaction of the patients/parents will be analyzed. Using a patient related expectation measurements (PREM) pre-operative expectations are investigated. Using a patient related outcome measurements (PROM) post-operative satisfaction is investigated. Children will be asked questions on teasing.
When the patient is younger than 12 years old, or mentally retarded, parents will fill in the questionnaire. When the patient is capable of completing the questionnaires we request the patient to fill in the questionnaires himself/herself.

For measurement of the facial/peri-orbital proportions of patients with syndromal craniosynostosis (Crouzon, Apert) and CFNS without hypertelorism (Control population I), distances on photographs will be measured using Photoshop (2D photographs) or Maxilim (3D-photographs). From a frontal view the midface is divided in 5 regions: the interocular distance, the ocular expanses and the distances between the lateral canthus to the medial part of ear.
We will evaluate the osseous proportions with CT/CBCT. Analysis will be done in Maxilim (CT, CBCT) and outcome will be described in millimetres and/or ratios. This will be done in Control population I (if these are available) and II. With this method, the height and width of the orbit will be measured, and the interdacryon distance determined in millimetres. All these measurements have been performed in Study population I and IIA pre-operatively and postoperatively. Then comparison with aged matched controls with syndromal craniosynostosis (Crouzon, Apert) and CFNS without hypertelorism (Control

population I) and with healthy peers (Control population II) can be done.

# Study description

### Background summary

Congenital craniofacial malformations are rare and cover a wide range of inborn anomalies of the skull and face. These deformities of osseous structures can cause both functional and esthetic problems that require surgical correction. This is certainly the case in deformities of the osseous structures in the peri-orbital zone, since visual functions and appearance of one of the most prominent facial features can be impaired. Peri-orbital osseous deformities occur in multiple congenital craniofacial disorders including 1. syndromic craniosynostosis, 2. midline facial clefts, 3. oblique facial clefts (uni- or bilateral), and 4. frontofacial (meningo-) encephaloceles. Major features of these disorders are hypertelorism, defined as an increased distance between the orbits; vertical (orbital) dystopia, meaning a discrepancy in the vertical position of both orbits; with or without midface hypoplasia, in which both the bones and soft tissues in the mid-portion of the face are underdeveloped in three dimensions. They all influence the position of the eye and the eyelids. The consequences of these features are inability to develop binocular sight, diplopia in all or parts of the visual field, incomplete closure of the eyelids and exposure keratitis. Patients with midface hypoplasia might also have a compromised airway with obstructive sleep apnea (OSA) and malocclusion. Next to these functional deficits there is also the aspect of looking different, with all its psychosocial impact.
Depending on the deformity and the anatomical structures involved, surgical techniques to correct the deformities include orbital box osteotomy, facial bipartition, monobloc advancement, and the several Le Fort procedures. However, the choice for the type of surgery and, particularly, timing of the surgical correction to achieve ideal facial proportions in relation to optimal function (eg. vision, intra-cranial pressure, occlusion, breathing) and growth remains controversial. These congenital deformities go hand in hand with a reduced growth potential of the affected areas, and following surgery growth potential might diminish as well. This means that a good result of an early intervention can deteriorate into a poor outcome over time.
Therefore, in this study the main objectives are better understanding of the effects of the various types of surgery, in relation to natural (restricted) growth in various pathologies. Normal growth in the normal population will be analyzed. This will aid in the decision making for the optimal type and timing of surgery for specific conditions. Improvement of the individualized management of patients with the different pathologies is our goal.

### Study objective

Primary objective:
To determine the most optimal technique and timing of surgery for correction of peri-orbital osseous deformities (hypertelorism, vertical orbital dystopia, and/or midface hypoplasia) for each specific condition (1. syndromic craniosynostosis, 2. midline facial clefts, 3. oblique facial clefts, and 4. frontofacial (meningo-)encephalocele). To achieve this goal we will:
a. Assess the functional and esthetic outcome and growth patterns in the peri-orbital region before and after correction, and relate this to technique and timing of surgery.
b. Compare these results with the results of patients suffering from the same condition that have not (yet) been operated for their peri-orbital osseous deformities, and a normal age-matched population without craniofacial malformations.

Secondary objectives:
2a. To determine expectations of surgery and satisfaction with results:
i. In surgeons: evaluate what they aim for as facial proportions of the upper face in each pathology, and assess whether they are satisfied with the results.
ii. In patients and parents: evaluate what their expectations are pre-operatively, and
what their satisfaction is postoperatively. In addition psychological evaluation is performed.
2b. To determine what facial/orbital proportions the surgeon should aim for in the correction of hypertelorism in patients with syndromal craniosynostosis (Crouzon, Apert) and craniofrontonasal syndrome (CFNS):
Assess whether facial/peri-orbital proportions of patients with syndromal craniosynostosis (Crouzon, Apert) and CFNS operated for hypertelorism, resemble proportions of syndromal craniosynostosis patients without hypertelorism (Crouzon, Apert), and CFNS and proportions of the normal population.

## Study design

This study can be categorised as an observational longitudinal study (according to the CCMO guidelines). The study uses a combination of a retrospective and prospective case-control and cross-sectional study design. Usually patients are followed until they reach the age of 21 years. The first evaluation of the prospective study will occur after 3 years.

## Study burden and risks

Regular care for these patients is carried out following an extended protocol. To investigate the objectives, the examinations we want to add to the existing protocol are two (3D-)photograph, and four questionnaires. We expect no associated risk. There will be no immediate benefit for the patient, but it will gain more insight in these pathologies and in the future this will lead to better patient care.

# Contacts

**Public**
Selecteer

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**Scientific**
Selecteer

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# Trial sites

## Listed location countries

Netherlands

# Eligibility criteria

**Age**
Adolescents (12-15 years)
Adolescents (16-17 years)
Adults (18-64 years)
Children (2-11 years)
Elderly (65 years and older)

## Inclusion criteria

Patients suffering from 1. syndromic craniosynostosis, 2. midline facial clefts, 3. oblique facial clefts (uni- or bilateral), and 4. frontofacial (meningo-) encephaloceles.

## Exclusion criteria

Patients are excluded if surgical correction was performed elsewhere or if data are incomplete, if patients in the Control group had peri-orbital surgical correction. Patients and parents do not speak the native Dutch language or are mentally retarded, will be excluded

from the questionnaires.

# Study design

## Design

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| Study type: | Observational non invasive |
| Intervention model: | Other |
| Allocation: | Non-randomized controlled trial |
| Masking: | Open (masking not used) |
| Control: | Active |
| Primary purpose: | Treatment |

## Recruitment

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| Recruitment status: | Will not start |
| Enrollment: | 500 |
| Type: | Anticipated |

# Ethics review

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| Approved WMO Date: | 21-01-2016 |
| Application type: | First submission |
| Review commission: | METC Erasmus MC, Universitair Medisch Centrum Rotterdam (Rotterdam) |

# 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 |
| --- | --- |
| CCMO | NL54142.078.15 |