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Orbital aspects following monobloc advancement in syndromic craniosynostosis☆

Céline A. Bender [a,*], Wouter Veneman [a], Jifke F. Veenland [b,c], Irene M.J. Mathijssen [d], Wim C.J. Hop [e], Maarten J. Koudstaal [a], Eppo B. Wolvius [a]

[a] Department of Oral and Maxillofacial Surgery, Erasmus University Medical Center Rotterdam, Netherlands
[b] Department of Medical Informatics, Erasmus University Medical Center Rotterdam, Netherlands
[c] Department of Radiology, Erasmus University Medical Center Rotterdam, Netherlands
[d] Department of Plastic Surgery, Erasmus University Medical Center Rotterdam, Netherlands
[e] Department of Biostatistics, Erasmus University Medical Center Rotterdam, Netherlands

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ABSTRACT

The monobloc advancement with distraction is a treatment modality for syndromic craniosynostosis, to correct exorbitism, upper airway compromise and malocclusion. In this report orbital volume and movements of (peri-)orbital structures and globes of seven patients following monobloc distraction are evaluated. In preoperative and postoperative CT-scans orbital volume was assessed and a 3D coordinate system with eleven landmarks was used to measure the movements of orbital structures and globes and to measure the change of exorbitism. Correlation between orbital volume, movements of the orbital structures and change in exorbitism was studied. The orbital volume increased by 49.9% (left) and 50.4% (right). The average anterior movement of the bone was 13.6 mm (left) and 13.9 mm (right). The mean anterior movement of the left globe was 5.8 mm and of the right globe 5.3 mm. The ratio of globe/bone movement was 0.4. Exorbitism decreased with 7.8 mm (left) and 8.1 mm (right). Differences between left and right for orbital volume, for movements and for the decrease in exorbitism were not significant. Volume enlargement and decrease in exorbitism were correlated ($p \leq 0.05$). Following monobloc advancement orbital volume increases, (peri-)orbital structures and the globe move forward and exorbitism diminishes.

1. Introduction

Craniosynostosis is a congenital disorder which is characterized by premature closure of the calvarial sutures and, which results in restriction of normal growth of the skull, brain and face. As a result patients suffer from a deformed skull and midface hypoplasia including shallow orbits (Gray et al., 2005; Harb and Kran, 2005; Khong et al., 2006; Cruz et al., 2007). These features can result in raised intracranial pressure, obstructive sleep apnoea and exorbitism (Ahmad et al., 2012). Therefore regular multidisciplinary screening is required, especially in children with Apert, Crouzon or Pfeiffer syndrome, in order to detect functional problems that indicate early surgical intervention promptly (Nout et al., 2008; Witherow et al., 2008a, b; Ahmad et al., 2012; Nout et al., 2012a).

A possible treatment is monobloc advancement, with or without distraction. The treatment is aimed at enlargement of the skull volume and advancement of the midface to correct raised intracranial pressure, improve upper airway obstruction and correct the exorbitism. As a secondary result, the patient's appearance will become less stigmatic (Ortiz-Monasterio et al., 1978; Witherow et al., 2008a; Ahmad et al., 2012).

Few articles report orbital changes following monobloc advancement (Cruz et al., 2007, 2008; Fitzgerald O'Connor et al., 2009; Ahmad et al., 2012). Fitzgerald et al. ($n = 10$) found that after monobloc advancement the nasion and the maxilla move forward (nasion: 12.9 mm; maxilla: 14.2 mm) as well as the globes (left globe: 5.3 mm; right globe: 6.3 mm) (Fitzgerald O'Connor et al., 2009). Ahmad et al. ($n = 12$) found that after monobloc

☆ The work should be attributed: Department of Oral and Maxillofacial Surgery (Head: Prof. Dr. E.B. Wolvius), Erasmus University Medical Center Rotterdam, Sophia's Children's Hospital, 's-Gravendijkwal 230, 3015 CE Rotterdam, Netherlands.

* Corresponding author. Department of Oral and Maxillofacial Surgery and Special Dental Care, Erasmus University Medical Center Rotterdam, Sophia's Children's Hospital, 's-Gravendijkwal 230, 3015 CE Rotterdam, Netherlands. Tel.: +31 10 7034138; fax: +31 10 7033098.
E-mail address: c.bender@erasmusmc.nl (C.A. Bender).

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advancement in eleven patients exorbitism diminished and protrusion of the eye decreased (from 154% to 109%) (Ahmad et al., 2012). Cruz et al. ($n = 38$) reported exorbitism to be corrected following monobloc advancement and the effect on both eyes to be symmetric (Cruz et al., 2007). Orbital volume changes, following monobloc advancement have not been reported.

In this paper, the relation between orbital volume enlargement, movements of the bony structures and the globes, and the change in exorbitism are analyzed by comparing preoperative and postoperative CT-scans in patients with syndromic craniosynostosis who underwent monobloc advancement.

2. Material and methods

2.1. Patients

Patients with syndromic craniosynostosis who underwent monobloc advancement between 2006 and 2010 were included. Preoperative and postoperative computed tomography scans (CT-scans) had to be available.

CT-scans were made in a supine position using the Emotion 6 (Siemens, Munich, Germany). Sedation was used when indicated. Slice thickness ranged from 1.0 to 2.5 mm and correction for different properties was carried out in the software program MeVisLab (Version 2.0, MeVis Medical Solutions AG, Bremen, Germany).

2.2. Surgical procedure

After a frontal craniotomy a 10-mm segment of frontal bone is left above the superior orbital rim. An orbital roof osteotomy is performed posteriorly to the midpoint of the globe, extending medially to the midline but with the anterior-most osteotomy line in front of the cribriform plate. The inferior orbital wall osteotomies are all made from above. Osteotomies are performed transversely through the nasal bone, caudally to the frontonasal suture and posteriorly to the lacrimal crest. The lateral osteotomies are made through the zygoma at the level of the frontozygomatic suture and more superiorly along the orbital rim. The midline osteotomy is directed inferiorly and posteriorly through the ethmoid. After pterygomaxillary disjunction Rowe disimpaction forceps are used to downfracture the maxilla. The distraction devices are placed on the zygoma, superiorly to the zygomatic arch in a posterior position. Once both devices have been placed, adequate mobilization is tested. At this point the stainless steel wires for external distraction are placed paranasally and at the bandeau. The frontal bone may be reshaped and angled posteriorly with segmental division of bone to achieve proper contour. The anterior cranial base is covered by a pericranial flap, which is secured to the surrounding cranial base bone in order to separate the nasal cavity from the dura. After closure of the cranial flap, the external frame is positioned with at least 4 transcutaneous pins on either side. Finally, the wires are fixed to the frame. After a consolidation period of 5 days active distraction is started with 5 mm per day.

2.3. Reproducibility

Measurements were independently repeated in all patients to assess the intraobserver reproducibility. Interobserver reproducibility was assessed for the movements of orbital structures.

2.4. Orbital volume

MeVisLab was used to analyze orbital volume in preoperative and postoperative CT-scans. In sagittal slices the anterior orbital boundary was determined as a straight line from the supraorbital rim to the infraorbital rim (Fig. 1). In case of bony interruptions (apertures or the optic nerve after leaving the curves of the orbital cone) a straight line between the two most approximate bony structures was drawn. These margins resulted in left and right orbital masks from which orbital volumes were calculated. A threshold of 400 Hounsfield units for bony structures was used to facilitate the segmentation.

2.5. Movements of orbital structures

In this same program (MeVisLab) the movements of the left and right globe, infraorbital rim, infraorbital foramen, lateral orbital rim, supraorbital rim and nasion were analyzed. To quantify these movements a coordinate system based on three planes in the preoperative CT-scan was computed as described in the study of Nout et al. (Nout et al., 2012b). Three reference points defined the horizontal plane, namely the most lateral points on the lateral semicircular canals and the anterior nasal spine. The vertical plane was defined by the left and right lateral semicircular canals and oriented at a 90-degree angle to the horizontal plane. The sagittal plane was oriented perpendicular to the horizontal and vertical plane. The sella turcica was taken as reference point and defined as (0, 0, 0) expressed in x, y and z coordinates. The postoperative CT-scan was rigidly registered to the preoperative CT-scan (Figs. 2 and 3).

Eleven landmarks were positioned independently in preoperative and postoperative CT-scans. The globe was defined by its midpoint in the axial, coronal and sagittal slices. Landmarks of the infraorbital and supraorbital rim were placed in that sagittal slice where the landmark of the globe was identified. The lateral orbital rim was defined in the axial slice where the midpoint of the globe was identified. Based on axial, coronal and sagittal slices the infraorbital foramen and nasion were positioned (Figs. 4 and 5). By comparing the coordinates preoperatively and postoperatively, the movement of the landmarks could be computed in three dimensions.

2.6. Exorbitism

The amount of exorbitism was defined as the perpendicular difference between the above-mentioned midpoint of the globes and lateral orbital rims.

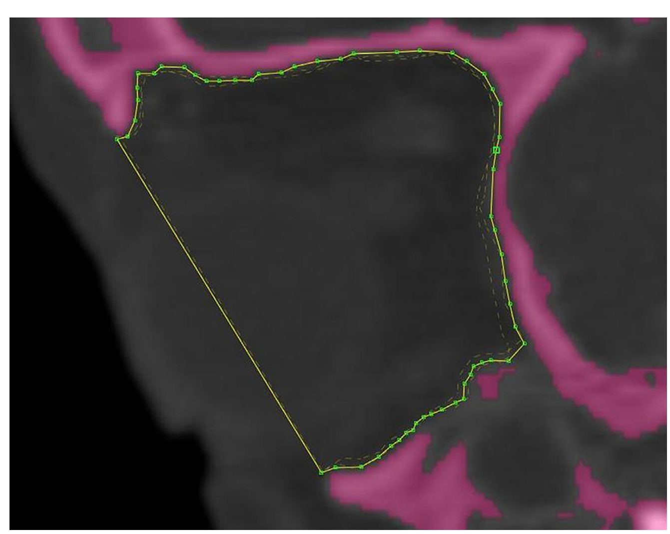

Fig. 1. The orbital boundary in a sagittal slice.