dition and the aesthetics concerns and delay the fronto-orbital advancement in syndromic craniosynostosis. Conventional techniques of posterior advancement are limited by scalp closure, supine head position in younger patients,slow brain expansion and bone quality.The use of distraction osteogenesis in the posterior vault,open a new frontier in the treatment of syndromic patients.The use of resorbable distractors apport the adventage of second intervention to remove the devices is not neccesary.

Methods and material: Retrospective evaluation of 6 cases of craniosynostosis syndromic patients (3 Crouzon, 2 Apert and 1 Saethre-Chotzen), operated by posterior cranial vault expansion with distraction osteogenesis as a first procedure.

Results: Volumetric pre and postoperative data showed an increase in posterior fossa and skull volumen in all the cases, improve the Chiari condition in almost 50% of the cases and improve the intracranial hypertension condition. No major complications or losses of the devices were present.

Conclusions: Posterior cranial vault expansion is an effective method to improve the intracranial hypertension tipically founded in syndromic craniosynostosis. The resorbables distractors are a safe, predectible and with low rate of complications materials for this technique.

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Spheno-orbital paediatric encephalocele and neurofibromatosis type 1. Custom-made skull base reconstruction

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Background: Sphenoid dysplasia is a manifestation of Neurofibromatosis type 1 (NF1), and occurs in over 7% of patients. Herniation of the temporal lobe through skull base defect produces spheno-orbialencephalocele. Surgical goals are vision and extraocular muscle preservation, as well as aesthetic condition improvement. Orbital anatomy reconstruction requires a combination of bone grafts, osteosynthesis and titanium meshes. Success and stability depend on the correct planning, reabsorption rate, brain pushing and neurofibromas growth.

Method: We present two cases of NF1 and spheno-orbital encephalocele, treated between 2012 and 2013. One and 2-year-old males presented with craniofacial deformity, dystopia and pulsating proptosis. Preoperative evaluation includes ophthalmologic assessment; computerized tomography and magnetic resonance. Studies demonstrate major sphenoid wing dysplasia; middle cranial fossa enlargement and in addition a plexiform neurofibroma of the III cranial nerve was also diagnosed. Planning starts with tridimensional reconstruction of DICOM images with the software Materialise, virtual model is generated with a specular orbital cavity to the non-affected side. Afterwards a stereolithographic (STL) model is fabricated. By means of an intracranial and lateral orbit approach, temporal lobe is retracted and bone defect expose. Over the STL model, the Titanium mesh is adapted, a custom-made graft is achieved for sphenoidal defect reconstruction.

Results: Follow-up period is longer than 6 months. Progressive craniofacial deformity improvement was achieved, besides proptosis recovery. Neither patient developed any important perioperative complications, and the postoperative radiological tests showed a complete encephalocele resolution.

Conclusion: Skull base reconstruction in sphenoid wing dysplasia can be accurately accomplished by computer-aided design technology. It is a safe procedure that decreases incorrect mesh adaptation and mobility, bone resorption, and encephalocele relapse.

Key words: encephalocele; orbital; neurofibromatosis; computer-aided design

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Airway management in neonates with robin sequence: the role of early mandibular distraction osteogenesis

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Background and objectives: Robin sequence (RS) is associated with glossoptosis, micrognathia, and airway obstruction. A U-shaped cleft palate may or may not be present. The retropositioned mandible pushes the tongue posteriorly and as such obstructs the airway, especially during feeding. In extreme cases surgical interventions such as tracheostomy and gastrostomy have been utilized. Alternatively, distraction osteogenesis (DO) relieves both airway obstruction and feeding difficulties by lengthening the jaw. The objective of this study was to evaluate the effectiveness of DO in RS.

Methods: A review of the current literature was performed to study the effectiveness of DO in managing airway and feeding problems in neonates with RS.

Results: In over 80% of the patients, conservative measures such as prone positioning and nasopharyngeal airway provides relief. In children who fail conservative therapy, DO provided resolution of airway obstruction and obviates the need of tracheotomy or allowed early decannulation, by increasing posterior airway space. In addition DO, resolved GERD and OSA severity in RS patients.

Conclusion: The underlying cause of airway obstruction and feeding difficulties in Robin sequence can be safely and effectively managed with DO. DO provide long-term improved outcomes with limited complications.

Key words: Robin sequence; distraction osteogenesis; airway

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