

Craniofacial Disorders and Surgical Treatment

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Abstract

In children with craniofacial problems, upper airway blockage can be treated using a variety of surgical and maxillofacial techniques. The nasopharyngeal prong is a straightforward concept that allows the airway blockage brought on by the tongue's posterior placement as a result of a small mandible to be swiftly and easily resolved without the need for more invasive surgical treatments.

Keywords: Temporomandibular joint • Temporomandibular joint disorders • Craniofacial abnormalities • Robin sequence

Introduction

The Pierre Robin sequence is a human craniofacial condition caused by non-coding mutations at the extreme end of a significant gene desert surrounding the SOX9 gene (PRS). We find two clusters of enhancers inside the PRS-associated area that control SOX9 expression during a constrained window of face progenitor development at distances up to 1.45 Mb using a human stem cell differentiation model. The Coordinator motif is required for the highly synergistic action seen by the enhancers in the 1.45 Mb cluster. We show using mice models that the two pathways that constrain Sox9 dosage perturbation to developing face tissues through context-specific enhancer activity and increase the susceptibility of the lower jaw to Sox9 expression decrease lead to PRS phenotypic specificity. Finally, we directly show that PRS is an enhanceropathy, identify the longest-range human enhancers implicated in congenital deformities, and show how even tiny variations in gene expression may cause morphological variance.

Description

Despite being relatively uncommon, craniosynostosis is frequently found in neurosurgical practises, particularly in paediatric neurosurgery. Sutural anatomy, its changes, and its effects on the brain, CSF, and the vascular supply, particularly the venous architecture of the skull and the dural sinuses, must be understood in order to manage this condition.

In an effort to enhance the comprehensive care of the craniosynostotic patient, the Journal of Pediatric Neurology and Medicine feels privileged to bring together the full spectrum of craniosynostosis with a variety of topics written by experts from around the world and India in this field. These topics range from a paediatric neurology perspective to advanced imaging and surgical techniques. We make a sincere effort to invite specialists in this area from all over the world to exchange information and insight. We owe the renowned authors a huge debt of gratitude for their contributions to our effort to improve and spread knowledge. A team of specialists, including paediatric neurosurgeons, plastic surgeons, maxillofacial surgeons, ophthalmologists, competent neuroanesthesiologists, qualified nurse practitioners, and

geneticists, perform craniosynostosis surgery. To optimise surgical correction and result, it is necessary to examine the secondary dynamic changes in more depth, including brain development, CSF circulation, and venous structure. A customised treatment protocol involving posterior calvarial augmentation, frontoorbital advancement, endoscopic third ventriculostomy, or ventriculoperitoneal shunt, either alone or in combination, is required for surgical planning. This includes thorough preoperative checks involving 3D imaging and reconstruction. Due to the extremely specialised examination and treatment techniques, very few neurosurgeons are well-versed in the extensive body of research on craniosynostosis.

The majority of neurosurgeons are more at ease treating single suture synostosis. Recent developments include endoscopy and the comprehensive management of syndromic craniosynostosis including Chiari I malformation and hydrocephalus, ophthalmological care in craniofacial disorders and hypertelorism and maxillofacial deformities, as well as imaging and surgical techniques including gradient echo gradient echo backbone and zero time echo MRI sequences, application of 3D photography in craniofacial surgery with future application in diagnostics using machine learning. The department of neurosurgery's specialised interdisciplinary craniofacial surgery units are primarily responsible for treating this. This broad and thorough collection of papers contributed by all leading specialists in this subject combines outstanding quality and substance for the benefit of our visitors. The craniosynostosis special issue covers a range of its aspects. It goes into great depth about both syndromic and nonsyndromic craniosynostosis across the board. The dialogues are open-ended, which gives our readers in this magazine varied perspectives.

Our eminent guest editors for the Journal of Pediatric Neurology and Medicine special issue, for their tireless efforts to get invited submissions from professionals in this area from India and throughout the world. The writers carefully selected the subtopics and demonstrated their depth of knowledge in their respective fields. This will undoubtedly be a reliable source of in-depth knowledge about craniosynostosis with the most recent developments in the area. It will encourage more study, advance our understanding of the topic, and help all people affected by craniosynostosis. This special edition is dedicated to the patients and their family, particularly those who have syndromic affection and whose fortitude and optimism are constantly tested. I hope that this supplemental information will serve as a resource for residents, junior neurosurgeons, and senior neurosurgeons to improve their understanding of craniosynostosis. In my opinion, this supplemental material serves as a fast reference for future investigation and study of a complicated and difficult subject like craniosynostosis.

Upper airway obstruction is one of the main causes of morbidity and death in newborns with craniofacial abnormalities. Babies that have craniofacial defects, such as Pierre Robin sequence, are at a higher risk of developing OSAS. These newborns are often monitored by a multidisciplinary team because to the complexity of their care, ensuring prompt evaluation and effective treatment. Clinical assessment may also involve genetic testing, imaging, endoscopy, and polysomnography in addition to the history and

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physical examination. Surgical and non-surgical treatment methods are available, and the choice will rely on the clinical evaluation, underlying condition, and degree of the disease. Although recent developments have improved the diagnosis and care of these individuals, numerous unanswered problems still exist. The existing research on the assessment and treatment of upper airway obstruction in neonates with craniofacial abnormalities is outlined in this review, with an emphasis on the Pierre Robin sequence [1-5].

Conclusion

A link between paediatric sleep disordered breathing and craniofacial discord is statistically supported. However, a marginally clinically significant increase in ANB angle of less than 2° in children with primary snoring and obstructive sleep apnea compared to controls should be considered. Therefore, this meta-analysis does not provide evidence for a direct causal link between craniofacial anatomy and paediatric sleep disordered breathing. Reduced upper airway width in kids with obstructive sleep apnea is strongly supported. The link between craniofacial and upper airway morphology and paediatric sleep-disordered breathing in all 3 dimensions needs to be examined in larger, well-controlled studies.

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Conflict of Interest

The author shows no conflict of interest towards this manuscript.

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