SURGICAL MANAGEMENT AND OUTCOME OF ISOLATED, NONSYNDROMIC SAGITTAL SUTURE CRANIOSYNOSTOSIS

Jontie Honey[1], Astrid Witters[2], Eline Misane MD[3], Jort van Rij[4]

[1]Master Student Medicine, University of Cambridge, Cambridgeshire, United Kingdom, [2]Master Student Medicine, University of Latvia, Riga, Latvia, [3]Erasmus Medical Centre, Rotterdam, The Netherlands, [4]Master Student Medicine, Maastricht University Centre, Maastricht, The Netherlands

Abstract

Review

Craniosynostosis jeopardises neonatal development both neurologically and physically. Surgical intervention is the main treatment strategy, though the method and timing of this surgery varies depending on the child's specific form of craniosynostosis and the parents/doctors' personal preference. To date, there is no internationally agreed consensus on the safest and most aesthetically efficacious method. In this article we summarise the pathophysiology of the disease, with a brief mention of the current understanding of the aetiology, to thus provide the basis for comparing the three main surgical intervention methods: spring-assisted cranioplasty (SAC), cranial vault remodelling (CVR) and strip craniectomy (SC).

Introduction

Craniosynostosis (cranio (skull); syn (together); ostosis (bone)), first described by Otto in 1830, is when one or more of the fibrous sutures in a neonate's skull ossifies prematurely. This has consequences for cranial and cephalic development by limiting the expansion perpendicular to the fused suture. You may think of the condition as an expanding balloon (the cranial vault with growing brain) that has been pinched at a specific section of the material so that expansion can only occur around this hindrance. It may be an isolated defect or a symptom of a syndrome – for example the Apert, Crouzon or Pfeiffer syndromes. Craniosynostosis on average presents in 1 in 2000 to 2500 births worldwide, of which 75% of the cases are boys. Saggital/scaphocephalic synostosis is the most common of nonsyndromic cases (50%), then coronal/anterior palgiocephalic (25%), metopic/trigonocephalic (10%), complex (10%) and lambdoid/posterior palgiocephalic (5%) (Figure 1,2) [1,2]. The prognosis of craniosynostosis is often bleak without surgical intervention. The "expanding brain in a rigid skull" means that there are almost always abnormalities in head shape and facial features as the unaffected suture regions undergo compensatory overgrowth to preserve the volume needed for normal brain growth. More severely, it may result in brain underdevelopment and cranial hypertension, which in turn may cause mental retardation, visual impairment, obstructive sleep apnea, and the Chiari malformation: the four most severe complications [3]. In this literature review we aim to compare the three main treatment forms, spring-assisted cranioplasty (SAC), cranial vault remodelling (CVR), and strip craniectomy (SC), using isolated, nonsyndromic sagittal synostosis as our case study. This is significant as these traditional treatment approaches involve surgical remodelling of the cranium and face, which carry significant morbidity and mortality risks. The future of craniosynostosis care looks to rely upon optimising these operations and on more effective screening and prenatal treatment.

Pathophysiology

To appreciate the pathophysiology, we must first understand the normal physiology. In normal skull development, ossification of the cranial vault commences around day 25 of gestation in the centre of each cranial bone, extending outward to the cranial suture [4]. The cranium is formed of eight bones separated by sutures: the sagittal suture to separate the two parietal bones; the coronal to separate the two frontal bones from the parietal bones; the metopic to separate the two frontal bones; the lambdoid separating the occipital bone from the two parietal bones.

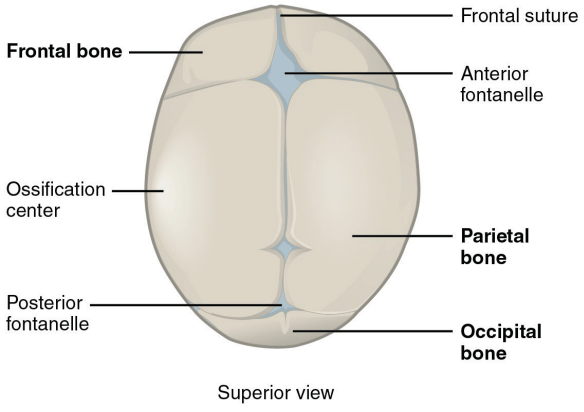

Figure 1: *Normal skull of the newborn*

Normal expansion occurs perpendicular to each suture: the anatomy allows expansion to be distributed across the entire skull. The sutures of the skull grow in response to tension within them generated by intracranial pressure. Thus, the primary factor that keeps sutures open is ongoing brain growth - as illustrated by the synonymously small cranium that is symptomatic of microcephaly.

If a suture fuses before 12 months of age, but typically before birth (craniosynostosis), skull growth is restricted perpendicular to the affected suture before the cranium has finished growing. In order to accommodate the growing brain, compensatory skull growth occurs parallel to the affected suture. The resulting skull deformity is dependent upon which suture(s) is/are affected. Scaphocephaly (from the Latin scaphoid, meaning boat) is the deformity where the sagittal suture is solely affected, resulting in restricted lateral expansion and compensatory anteroposterior growth, namely growth at the coronal and lambdoid sutures. The neonate presents with frontal bossing and occipital coning. This is the most common craniosynostosis.

The aetiology of isolated, nonsyndromic craniosynostosis is still to be elucidated, though the associated syndromes provide some light on possible mutational causes: most syndromes show mutations in the genes that code fibroblast growth factor receptor (FGFR) [5]. Though management is mostly focused on surgical remodelling of the neonate's skull, it is important to note the future potential therapeutic interventions using prenatal/intrauterine methods based on evolving molecular and genetic

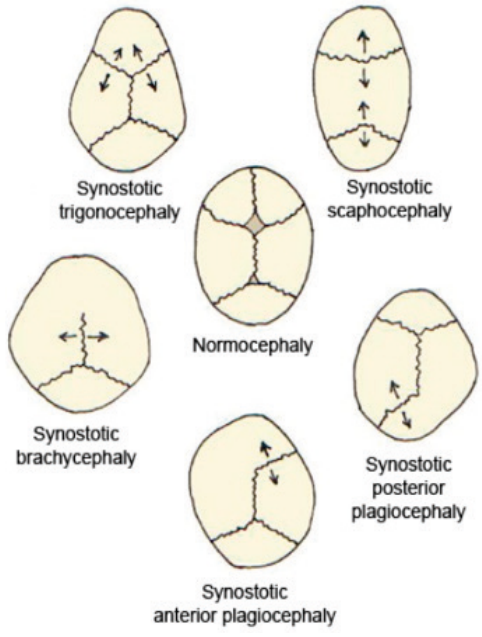

Figure 2: *Different types of craniosynostosis*

understanding (Wan DC, 2008). Three of the four FGFRs have been associated with premature pathologic suture fusions, providing a doorway for pharmacotherapeutics.

Surgical management

Surgical interventions for sagittal suture craniosynostosis aim to sufficiently remodel calvarial shape for brain development, reduce intracranial pressure (ICP), and improve aesthetic appearance. The primary aesthetic measure of most studies concerning surgical intervention for scaphocephaly is the cephalic index (CI) – the biparietal diameter (width) of the head multiplied by 100 and divided by its occipitofrontal diameter (length). To date there are three main treatment forms: spring-assisted cranioplasty (SAC), cranial vault remodelling (CVR), and strip craniectomy (SC). SAC involves a small-scale craniectomy at the fusion, small osteotomies either side of the fused sagittal suture and the placement of metal springs that gradually widen the gap to encourage new osteogenesis between the two cut surfaces. CVR involves open surgery to temporarily remove the cranium (a craniotomy), reshaping of these removed bones, and reinsertion of these bones secured using synthetic plates and dissolving secures. CVR is the most invasive procedure. SC is performed by placing small incisions at both ends of the sagittal suture, through which the fused suture and approximately two inches of surrounding bone is removed. SC is the least invasive but requires the use of a moulding helmet to guide the bone growth.

With regard to timing, there is a distinction to operate before or between 6 and 12 months – almost all surgeries occur within the first year of life. Before six months the used techniques are the SAC or SC. After six months, more commonly an open CVR is performed. The best treatment of craniosynostosis would keep blood loss and recovery time to a minimum, whilst achieving the desired intracranial volume. Note that, unsurprisingly, when there is an associated syndrome involved the mortality and morbidity are higher because these children have an increased risk for complications and often a more challenging surgery [6].

Outcomes

The effectiveness of SAC has been compared to the modified pi-cranioplasty regarding morphological outcomes and procedure safety – there needs to be an increase in the baby's intracranial volume and the establishment a more "normal" craniofacial appearance. All three techniques achieve a relatively similar morphological outcome. The pi-plasty group has a CI slightly closer to the normal range at the age of 3, than after SAC. But regarding blood loss, transfusion requirements, operative time, ICU time, recovery time and total hospital stay the SAC group was superior compared to the pi-cranioplasty group [7]. Additionally, endoscope-assisted surgery is also commonly used in sagittal craniosynostosis surgery. Iyer et al. [8] concluded that a single incision technique improves the classical surgical procedure by decreasing invasiveness, reduces intraoperative blood loss, reduces surgery time and has a cosmetic advantage because only one incision must be made. After six months, the techniques above are limited in their efficacy. In this case CVR is performed.

Chummun et al. [9] compared CI after different types of CVR: the open calvarial, subtotal remodelling and the more conservative strip craniectomy. The results showed an improvement in the CI after all types of cranial vault remodelling. The open calvarial vault surgery resulted in a greater CI and a more mesocephalic shaped head. However, the age at which the surgery was performed varied considerably, so a selection bias cannot be excluded. Gerety et al. [10] demonstrate that CVR, SC and SAC provide adequate correction of CI in the short term. When CVR is compared with SMC, no significant difference in correction of CI was observed (weighted mean difference (WMD = 0.94 (95% CI: -0.23-2.11; I^2=55%, p = 0.12)). When compared with SC, CVR creates a small but significantly greater improvement in CI (WMD = 1.47 (95% CI: 0.47-2.48; I^2=66%, p = 0.004)). Average postoperative CI was correlated with average follow-up across techniques; this correlation demonstrated that as average follow-up increased, CI increased in the SC and SMC groups and decreased in the CVR group.

In a systematic review conducted by Maltese et al. [11], two out of the three studies involving the use of SAC did not show any difference in postoperative CI between the methods, while the third study showed

Table 1: *Comparisons of the different types of surgery*

	Spring-assisted Cranioplasty	Cranial Vault Remodelling	Strip Craniectomy
Performed	3-8 months of age	> 6 months of age	< 6 months of age
Procedure time	3-4 hours	5-6 hours	2-3 hours
Recovery time	2-3 days	4-7 days	2 days
Blood transfusion rates (% of total blood)	~50%	Highest (~100% blood transfusion)	~50%
Aesthetics (scarring, CI)	Sagittal scars; normal CI	Ear-to-ear scar; normal CI	Two scars on top of scalp (anterior and posterior); normal CI
Other notes	Will require a second surgery 8-12 weeks after in order to remove the springs		Helmet required

that SAC was slightly worse than the pi-plasty technique. Since the studies had a high risk of bias, it can be concluded that it is uncertain whether the SAC technique is comparable to alternative techniques regarding CI as outcome. However, it should be noted that the quality of evidence was very low.

Conclusion

The prognosis of craniosynostosis is often bleak without surgical intervention. Based on the literature, surgical intervention procures a better prognosis than no intervention. Nowadays there is a broad range of modern surgical techniques available to treat sagittal craniosynostosis. All the techniques reach an improvement in the CI and reduce ICP. The pi-plasty reaches a more 'normal' skull, the SAC and endoscope-assisted techniques score better on factors like blood loss. After six months, the CVR involves more complications but also reaches a CI enhancement.

Future research

Further research may focus on standardisation of the mean human skull, comparative surgical reduction of morbidity and generating international care guidelines, as presented by the working group of craniosynostosis from the Netherlands. This can be reached by performing an RCT where the different surgical techniques are compared and the CI is mapped as well as international collaboration into the neuro-cognitive development assessed with IQ postoperatively.

References

1. Lee HQ, Hutson JM, Wray AC, Lo PA, Chong DK, Holmes AD, et al. Changing epidemiology of nonsyndromic craniosynostosis and revisiting the risk factors. The Journal of craniofacial surgery. 2012;23(5):1245-51.
2. Cornelissen M, Ottelander B, Rizopoulos D, van der Hulst R, Mink van der Molen A, van der Horst C, et al. Increase of prevalence of craniosynostosis. Journal of cranio-maxillo-facial surgery : official publication of the European Association for Cranio-Maxillo-Facial Surgery. 2016;44(9):1273-9.
3. Renier D, Lajeunie E, Arnaud E, Marchac D. Management of craniosynostoses. Child's nervous system : ChNS : official journal of the International Society for Pediatric Neurosurgery. 2000;16(10-11):645-58.
4. Ursitti F, Fadda T, Papetti L, Pagnoni M, Nicita F, Iannetti G, et al. Evaluation and management of nonsyndromic craniosynostosis. Acta paediatrica (Oslo, Norway : 1992). 2011;100(9):1185-94.
5. Twigg SR, Wilkie AO. A Genetic-Pathophysiological Framework for Craniosynostosis. American journal of human genetics. 2015;97(3):359-77.
6. Sloan GM, Wells KC, Raffel C, McComb JG. Surgical treatment of craniosynostosis: outcome analysis of 250 consecutive patients. Pediatrics. 1997;100(1):E2.
7. Windh P, Davis C, Sanger C, Sahlin P, Lauritzen C. Spring-assisted cranioplasty vs pi-plasty for sagittal synostosis--a long term follow-up study. The Journal of craniofacial surgery. 2008;19(1):59-64.
8. Iyer RR, Uribe-Cardenas R, Ahn ES. Single incision endoscope-assisted surgery for sagittal craniosynostosis. Child's nervous system : C hNS : official journal of the International Society for Pediatric Neurosurgery. 2017;33(1):1-5.
9. Chummun S, McLean NR, Flapper WJ, David DJ. The Management of Nonsyndromic, Isolated Sagittal Synostosis. The Journal of craniofacial surgery. 2016;27(2):299-304.
10. Gerety PA, Basta MN, Fischer JP, Taylor JA. Operative Management of Nonsyndromic Sagittal Synostosis: A Head-to-Head Meta-analysis of Outcomes Comparing 3 Techniques. The Journal of craniofacial surgery. 2015;26(4):1251-7.
11. Maltese G, Fischer S, Strandell A, Tarnow P, Kolby L. Spring-assisted surgery in the treatment of sagittal synostosis: A systematic review. Journal of plastic surgery and hand surgery. 2015;49(3):177-82.
12. Wan DC, Kwan MD, Lorenz HP, Longaker MT. Current Treatment of craniosynostosis and future therapeutic directions.

INTERNATIONAL SUMMER SCHOOL NEUROSURGERY: BRIDGING GAPS

RAMS is and will always be about inspiring people, exchanging knowledge and creating engaging communities of aspiring academics. Every year, an omnifarious set of lectures, workshops and journals is presented to an audience of students and staff of Radboud University and the Radboud University Medical Center. A new chapter was started this summer, when RAMS organised its premiere major international event: the very first International Summer School in Neurosurgery, bridging gaps for the second year in a row after the successful first Nijmegen edition last year.

Students from all over Europe and beyond took part in a week full of engaging and motivating activities. Together with the staff of the Neurosurgical Centre Nijmegen (NCCN) and various national and international European collaborators, many lectures, discussions and workshops were organised by the Summer School Committee to emerge forty students in the field of neurosurgery. In addition to taking part in discussions covering future perspectives and state-of-the-art operating procedures, students discussed neuroanatomy in the dissection rooms of the medical faculty, performed surgery with professional Stryker equipment on 3D-printed crania from Delta Surgical and presented their own ideas and expectations of the field in front of a jury of neurosurgical representatives.

Bridging gaps is all about finding a fitting interdisciplinary approach, which the committee found in a collaboration with the Donders Institute for Brain, Cognition and Behaviour. This way, the Summer School not only covered neurosurgical practice, but students also discussed neuroscientific backgrounds and were encouraged to extrapolate research questions from the various activities.

The evaluation of the participants was very positive and will be of great help for RAMS' future international activities. RAMS has entered a new era of international activities and will continue its journey, giving voice to academic potential.

Special thanks to the Summer School Committee for making the International Summerschool unforgettable: Jules Janssen Daalen, Dirk Loeffen, Jill Martens, Barof Sanaan and Daan Viering.