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Spring Assisted Cranioplasty for the Correction of Non-Syndromic Scaphocephaly: A Quantitative Analysis of 100 consecutive cases

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Abstract

Background

Spring-assisted cranioplasty has been proposed as an alternative to total calvarial remodelling for sagittal craniosynostosis. Advantages include its minimally invasive nature, reduced morbidity and hospital stay. Potential drawbacks include the need for a second procedure for removal and the lack of published long-term follow-up. We present a single institution experience of 100 consecutive cases using a novel spring design.

Methods

All patients treated at our institution between April 2010 and September 2014 were evaluated retrospectively. Patients with isolated non-syndromic sagittal craniosynostosis were included. Data were collected for operative time, anaesthetic time, hospital stay, transfusion requirement and complications in addition to cephalic index pre-operatively and at one day, three weeks and six months post-operatively.

Results

One hundred patients were included. Mean cephalic index was 68 pre-operatively, 71 at day 1 and 72 at 3 weeks and 6 months post-operatively. Nine patients required transfusion. Two patients developed a CSF leak requiring intervention. One patient required early removal of springs due to infection. One patient had a wound dehiscence over the spring and 1 patient sustained a venous infarct with hemiplegia. Five patients required further calvarial remodelling surgery.

Conclusion

Our modified spring design and protocol represents an effective strategy in the management of single-suture sagittal craniosynostosis with reduced total operative time and blood loss when compared to alternative treatment strategies. In patients referred within the first 6 months of birth this technique has become our procedure of choice. In a minority of cases especially in the older age groups further remodelling surgery is required.
**Introduction**

Multiple procedures continue to be used for the surgical management of single-suture sagittal craniosynostosis globally, with no clear consensus on timing or nature of intervention. The operative strategies published can be divided into 3 major groups: total calvarial vault remodelling (TCR) in older children; a subtotal calvarial vault remodelling or a modification of the pi procedure in younger children; and “minimally invasive” techniques. Of the latter group, unenhanced simple strip craniectomy has largely been abandoned due to poor results.\(^1\) However, the potential benefits of other minimally invasive techniques, namely endoscopic strip craniectomy with helmet therapy or head banding and spring-assisted cranioplasty (SAC), have led to much interest. Minimally invasive techniques are attractive, particularly in young infants (< 6 months), as alternatives involve a long bicoronal scar, prolonged hospitalization, substantial blood loss and with a high chance of requiring allogenic transfusion requirements.\(^2\)–\(^5\)

The first SAC was reported by Lauritzen et al.\(^6\) The principle of SAC is to use the viscoelastic properties of the immature skull to widen the vault and reduce cranial length. Experimental evidence suggests that this process stimulates excess growth in the coronal and lambdoidal sutures, in a manner physiologically synonymous with distraction osteogenesis.\(^7\) Putative advantages of SAC include: a smaller incision; less soft tissue and dural dissection; a smaller number of osteotomies; reduced blood loss and transfusion; less risk of dural breach/brain injury; and shorter hospital stay. Criticisms of the technique have pointed out the need for a second, smaller, surgical procedure to remove the inserted springs, the possibility of undercorrection of the scaphocephaly, and the lack of long-term published follow-up. This operation is in evolution and there exists neither a standard surgical technique nor standard spring design.\(^8\) At Great Ormond Street Hospital for Children Craniofacial Unit, London, UK we have developed a minimal access method of SAC utilising our own
spring design. In this paper, we present the results of our first 100 consecutive cases using a standardised, refined approach.

**Patients and methods**

**Patient selection**

Spring assisted craniofacial procedures started at GOSH in January 2008 after an 8 month period of distractor design. Initially, posterior vault expansion procedures were undertaken utilising these springs. In April 2010 this technique was extended for use in scaphocephaly correction. To date, over 300 patients have undergone spring assisted cranioplasty procedures for which a contemporaneous database is maintained. The database was interrogated for all patients with non-syndromic sagittal craniosynostosis managed by SAC and the first 100 consecutive cases between April 2010 and September 2014 inclusive were included.

All patients referred with scaphocephaly are evaluated by a consultant craniofacial surgeon (plastic or neurosurgeon), and the diagnosis of isolated sagittal synostosis is made on the basis of clinical examination supplemented in some cases by radiological imaging (plain film radiograph or 3-dimensional CT reconstruction), when required for diagnostic and planning purposes. Once the diagnosis is established, the reviewing surgeon discusses the following options with the parents: for children of 6 months of age or less – conservative management, SAC or a modification of the pi procedure; for older children – conservative management or TCR. The prospective advantages and disadvantages of each management strategy are explained and the final decision made by the family. If, following clinical assessment, the reviewing surgeon felt SAC appropriate, it was offered to some children older than 6 months (though none over 12 months). Patients with multi-sutural or syndromic craniosynostosis, including those first thought to be non-syndromic but later identified as having a genetic
diagnosis, and those whose parents opted for TCR or the modified pi procedure were excluded from the analysis.

Operative technique

For SAC, the anaesthetised patient is positioned prone with the neck extended (“sphinx position”) see Figure 1, with the vertex parallel to the operating room floor. A single 8cm scalp incision is made, perpendicular to the sagittal suture, midway between the anterior and posterior fontanelles with adjustments made for the particular type of scaphocephaly and therefore the location of the springs to be inserted. Dissection proceeds along the sub-galeal plane to the anterior and posterior fontanelles and in their absence from the coronal to the lambdoid sutures. A 15mm square craniectomy is next performed straddling the sagittal sinus, approximately halfway along the fused sagittal suture. Following complete dissection of the dura away from the inner table in the line of the planned release, two parallel parasagittal osteotomies are made starting from the craniectomy site, extending to the coronal sutures anteriorly and to the lambdoid sutures posteriorly. These osteotomies are parallel to the sagittal suture for the most part fanning out a little posteriorly as they meet the lambdoid sutures. Occasionally midline bridging veins are encountered and these are easily controlled via the access created. At the end of the osteotomies, the midline bone struts should buckle with relative ease on the coronal or lambdoid sutures with which they remain connected. A central bar is maintained to aid in the ossification of the distracted cranium. Two GOSH springs (see spring design below) are then placed into prepared grooves, ensuring the spring footplate design is firmly locked. The spring strength is chosen by the operating surgeon dependent on on-table trials. Ideally an on table expansion on either side of the midline of 1cm should be expected. A drain is placed in the sub-galeal plane and the skin is closed in layers with resorbable sutures. Post-operatively, the child is nursed initially in a post-operative intensive recovery setting until fully recovered from general anaesthesia and then
returned to a general neurosurgical ward for observation. The wound drain is removed at 24 hours and a post-operative check X-ray is performed on the first post-operative day. The child is reviewed daily by the craniofacial team and discharged home when appropriate, typically on the first post-operative day. Spring removal is typically undertaken 3 months later (following maximal radiographic spring expansion and interpositional bone growth) through the same incision under general anaesthesia as a day-case procedure.

Spring design

The spring distractors used are stainless steel standardised wireform springs, bearing a central loop and angled hooks at the end of each leg, which slot into bony grooves fashioned following osteotomy. The aim is to use the simplest possible design that would allow precise measurement of in-vivo spring dynamics by means of medical imaging. The springs have a 6 cm unloaded opening and a gentle curvature to allow a better fit with the calvarium. There are three models with the same geometry but varying wire thickness of 1.0, 1.2 and 1.4 mm (named S10, S12 and S14) corresponding to increasing loading and unloading stiffness. These springs have been used at our institution since 2008. In figure 2, a sample of the crimping curve for a cranioplasty spring is reported.

Data acquisition

Data were collected from hospital records and included operative duration, total anaesthetic time, length of hospital stay, transfusion requirement, and complications. Outcome was measured using the cephalic index (the ratio of biparietal diameter to the occipito-frontal diameter multiplied by 100) derived from skull radiography pre-operatively and at day 1, 3 weeks and 6 months post-operatively. Inter-rater agreement was tested using Bland-Altman plots.
Statistical analysis

Preoperative cephalic index was compared to cephalic index measured at day 1, 3 weeks and 6 months post-operatively. Data were analysed using one-way analysis of variance (ANOVA) with Dunnett’s post test correction for multiple comparisons using the pre-operative cephalic index measurements as the control. Inter-rater agreement was analysed using Bland-Altman plots. Significance was set at P<0.05.

Results

Demographics and hospital data

100 patients were included in the study. There were 23 female and 77 male patients. The mean age at spring insertion was 169 days (±48 days, 1SD), and the mean age at removal was 272 ± 73 days, 1SD. The mean duration of expansion prior to spring removal was 104 ±50 days, 1SD. Mean operative time for spring insertion was 58 ±17 minutes, 1SD (114 ±21 minutes, 1SD including anaesthetic induction and recovery). The median duration of in-hospital stay for spring insertion was 33 hours (range 23 to 1332 hours). The mean operative time for spring removal was 37 ±13 minutes, 1SD (81 ±14 minutes, 1SD including anaesthetic induction and recovery). The median duration of in-hospital stay for spring removal was 10 hours (range 4-210 hours).

Cephalic index

Cephalic index (CI) was available for the 82 patients who had pre-operative imaging. 18 patients did not have any pre-operative radiographs and therefore CI could not be calculated. Post-operative CI was available for 95 patients at day 1 (suboptimal radiographs in 5 patients), for 91 patients at 3 weeks (radiographs were not available for 5, and were suboptimal in 2 patients), and for 23 patients at 6 months (radiographs were not present for 75 and suboptimal in 2). Concordance was assessed using a Bland-Altman plot, confirming an acceptable bias of 0.3 ± 0.9, 1SD. Mean CI was 67.7 ±3.6, 1SD pre-operatively, 71.0
±4.5, 1SD at day 1, 72.2 ±4.8, 1SD at 3 weeks and 72.1 ±3.5, 1SD at 6 months. ANOVA confirmed the alternative hypothesis that mean CI was different at one or more time points. The mean difference between pre- and day 1 post-operative CI was 3.3, and Dunnett’s post-test confirmed statistical significance (P<0.0001). The mean difference between pre- and week 3 post-operative CI was 4.5 (P<0.0001). The mean difference between pre- and 6 month post-operative CI was 4.4 (P<0.0001) (figure 3).

Transfusion Requirements

Of 100 SAC insertions, 92 patients did not require blood transfusion. The 8 patients transfused received a mean of 143 ml of allogenic blood. 5 patients had a single donor exposure and 3 patients had 2 donor exposures. For spring removal, 1 patient in 100 required a transfusion of 200 mls (1 donor exposure).

Over all 100 patients, 9 patients were transfused with a combined mean transfusion requirement for insertion and removal of 13.5 ml. For spring insertion there was a mean blood transfusion requirement of 11.5 ml (range 0-310 ml).

Complication profile

Complications were seen in 9 patients following spring insertion (Table 1). Grade 1 complications (defined as no delay in discharge, re-operation or longterm sequelae) were as follows: two patients had unintended durotomies which were repaired at the time of the procedure. Grade 2 complications (defined as a delay in discharge but no further operation required) were as follows: one patient developed a post-insertion sub-galeal haematoma that delayed discharge but did not require further intervention; one patient developed a localised infection and was admitted for observation and intravenous antibiotics several days prior to their planned spring removal. Grade 3 complications (defined as a re-operation but no longterm sequelae) were as follows: one patient had a spontaneous exposure of one spring seven days post-insertion due to wound dehiscence over the spring; one patient had springs
removed early due to surgical site infection; one patient developed a CSF leak requiring return to theatre for re-suturing of the wound; one patient developed a CSF leak requiring a lumbo peritoneal (LP) shunt and recovered uneventfully; no organisms were cultivated from the LP shunt following removal and the patient was well at discharge however, 2 months later the patient developed pneumococcal meningitis, believed to be community acquired, and subsequent hearing loss. This patient went on to have a 3cm bone defect over the vertex which required a split calvarioplasty 4 years following the initial procedure. Grade 4 complications (defined as longterm deficit or neurological impairment or permanent disability) were as follows: one patient developed intra-operative bleeding secondary to an iatrogenic injury to a large parasagittal vein resulting in a large sub-dural haematoma and subsequent venous infarct. The clot was removed promptly but the recovery was incomplete with residual hemiparesis (Grade 4 plus in the leg and grade 4 in the arm) and homonymous hemianopia. The surgical technique has since been adapted to improve safety related to the sagittal sinus and there have been no further events following this intervention (see discussion). With regards to the spring removal procedure, other than 1 child requiring transfusion (as detailed in the Transfusion Requirements section above), there were no surgical complications recorded.

Follow up
Mean length of follow up was 38 months (range 11 – 65 months). 95% of patients in this cohort have not required further craniofacial surgery within the follow up period studied. In our cohort 5 patients required further remodelling; in 2 patients the indication was raised ICP and in 3 patients the indication was residual head shape concern. Those patients requiring remodelling for raised ICP had posterior vault expansions 3 and 4 years post-operatively. Of those requiring remodelling for residual head shape concern 2 had fronto-orbital remodelling at our unit 2 years post-operatively and 1 had TCR at a different unit 4 years post-operatively.
Discussion

We report the largest series of SAC for isolated non-syndromic sagittal synostosis published to date that demonstrates that this minimally invasive technique is safe and well-tolerated, with low transfusion requirements and short inpatient hospital stays, and is effective in improving CI, at least within the study period. It has been adopted as a standard technique in our supraregional craniofacial service and is offered routinely as an option to parents of infants (under 6 months of age) presenting with scaphocephaly. There exists a wide variety of opinions with regards to the appropriate management of non-syndromic sagittal craniosynostosis.\(^9\) Our experience has been that, when the various treatment options for scaphocephaly are discussed with the parents, SAC is attractive due to its minimally invasive technique and reduced hospitalization when compared to traditional TCR. During the same study period 26 patients with isolated sagittal synostosis were treated with TCR, 12 with antero-posterior shortening +/- biparietal widening and 3 with anterior two thirds remodelling.

Even when considered as a “2 stage” procedure – i.e. SAC itself and then a subsequent admission for surgical removal of the springs, the combined numbers are favourable. Total operating time (95 minutes), total anaesthetic time (195 minutes), total hospital stay (43 hours) and mean transfusion requirement (13.5 ml) are all significantly less than published series of TCR.\(^2,10,11\) The reduced blood loss and physiological stress inherent in this minimally invasive technique have reduced the complexity of anaesthesia for these often very small infants. For example, these children no longer have arterial lines routinely sited, which were mandatory when our unit performed TCR as the standard early scaphocephaly correction. Our anaesthesia team are planning to report their experience as a separate publication.

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Reducing the number of neurosurgical bed days and theatre time required improves efficiency and allows other children to be treated in a timely manner, reducing waiting times for other craniofacial and neurosurgical admissions.

As we report, in those for whom radiographic data were available, SAC results in a significant and sustained improvement in CI. Our reported CI improvement is comparable to the literature,\(^\text{(8,12–15)}\) in particular with experience of traditional TCR techniques.\(^\text{(1–4)}\) In addition, CI improvement with SAC is similar to that achieved with the minimally invasive technique of helmet-assisted endoscopic sagittal strip craniectomy.\(^\text{(16–18)}\) Whilst change in CI is only one aspect of scaphocephaly correction, it is currently in standard use as an outcome measure for this type of surgical procedure.\(^\text{(19)}\) The deformity associated with isolated sagittal synostosis is more complex than a simple mismatch of anteroposterior length and biparietal width and includes frontal bossing; temporal indrawing (or “pinching”); an anteriorly placed vertex and protuberant occiput (“bullet”). To assess this complex 3D deformity, and the effect thereon of surgery, we are now performing on-table 3D optical photography before SAC, as well as at the time of spring removal to quantify our results in a more meaningful fashion (Figure 4). We plan to report these results in the near future.

Currently, the choice of which calibre (and therefore strength) spring to use is decided on a case-by-case basis by the operating surgeon dependent on on-table trials. As our clinical experience increases, we hope to use follow up radiographic, 3D morphometric and clinical outcome data, as well as laboratory-based bio-engineering studies, to help predict head shape outcome after SAC and aid in pre-operative planning, including choice of spring strength.

Concerns have previously been raised in the literature about possible undercorrection of the scaphocephalic deformity by minimally invasive techniques.\(^\text{(15)}\) In the current series, 5 children did require further corrective surgery; 3 because of ongoing appearance concerns regarding residual deformity (Figure 5) and 2 because of presentation with clinical evidence
of raised intracranial pressure. The decision for secondary remodeling surgery is made for aesthetic reasons following discussion with the parents and not on CI measurements alone. 95% of children did not require further surgery in the study period (Figure 6). Because of this finding, we do advise parents at the initial consultation that undercorrection may occur in a small proportion of children with isolated sagittal synostosis treated with SAC, and that this rate may be less in those managed with a traditional formal TCR. However, the small risk of undercorrection in SAC must be balanced against the advantages of minimally invasive surgery as discussed above. In addition, the role of early corrective surgery for prevention of raised intracranial pressure and/or improving neurodevelopmental outcome remains controversial however, the 2% incidence of raised intracranial pressure reported in this series compares favourably with the baseline rate in isolated sagittal synostosis, which has been reported to be as high as 44%.(20)

In our series there was one serious neurological event – a venous stroke resulting in hemiparesis. The severity of this complication led to a review of the technique. Analysis of that particular case revealed that an unintended dural breach had occurred underneath one of the osteotomies – damaging a parasagittal draining vein with resultant venous congestion and ischaemia. Up to this point, the osteotomies had been cut from burr holes fashioned 1 cm lateral to the fused sagittal suture. Following this complication, the technique was adapted – a 15 mm central craniectomy is performed in the midpoint of the fused suture. This allows dissection of the dura away from the inner table under direct vision to the full extent of the suture anteriorly and posteriorly, reducing the risk of inadvertent injury when cutting the osteotomy. This complication occurred within 18 months of the start of this study, and since modification of the technique we have not had a further neurological complication following SAC within the unit.
As with any novel technique, one disadvantage of such series is the lack of long-term follow up. Initial evidence presented in this report suggests a sustained improvement in CI over 1 year, however we are following up this cohort to assess robustness of correction into later childhood, adolescence and adulthood. Our now routine use of 3D optical imaging will support our ongoing clinical assessments, and we also plan to introduce patient-centred outcome measurements to report parental and child satisfaction with results of corrective surgery. We look forward to publishing these results in due course to share the long-term outcomes of SAC.

Overall, we believe that within the context of cranioplasty for non syndromic sagittal craniosynostosis SAC is a safe and effective minimally invasive technique for management. We do not advocate that SAC should replace traditional, formal TCR but rather complement it within the surgical “toolbox” of the complete craniofacial unit. Patient selection, including well-informed parental consent, remains paramount and it may not be the most appropriate operation for all children. We certainly believe, however, that SAC has a role to play in the management of scaphocephaly in infants.
References


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Legends

Figure 1: Operative technique (A) – demonstrating ‘sphinx’ positioning; (B) – showing location of scalp incision midway between anterior and posterior fontanelles with burr holes; (C) – showing site of 15mm square craniectomy; (D) – showing location and posterior fanning of parasagittal osteotomies with orientation of springs; (E) – showing direction of cranial expansion.

Figure 2: Sample of crimping curve of an S10 cranioplasty spring (shown on the right), with force values reported on the horizontal axis.

Figure 3: Box-and-whisker plot to show change in cephalic index. Mean CI was 67.7 ±3.6, 1SD pre-operatively, 71.0 ±4.5, 1SD at day 1, 72.2 ±4.8, 1SD at 3 weeks and 72.1 ±3.5, 1SD at 6 months (P<0.0001).

Table 1: Complications at Insertion (no complications recorded at removal)

Figure 4: Scans of a sagittal patient taken with a 3D handheld scanner before the insertion and after the removal of springs. The colour map shows surface distances between both scans.

Figure 5: From left to right Lateral, Frontal and Superior views at pre spring insertion (A); 2 months post removal of springs (B); 2 years post removal of springs and prior to anterior 2/3rds remodelling (C) and 10 months post anterior 2/3rds remodelling (D).

Figure 6: From left to right Lateral, Frontal and Superior views at pre spring insertion (A) and 3 months following removal of springs (B).
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<th>Type</th>
<th>Description</th>
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<td>Type I</td>
<td>No delay in discharge, re-operation or longterm sequelae</td>
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<td>Delay in discharge but no further operation required</td>
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<td>Re-operation but no longterm sequelae</td>
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Figure 2
Figure 3

A box plot showing the Cephalic Index over time:
- Pre-op
- Day 1
- 3 weeks
- 6 months

Significant differences indicated by ***
Figure 4
Figure 5
Figure 6