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Results following adoption of a modified "Melbourne technique" of total scaphocephaly correction at Great Ormond Street Hospital --Manuscript Draft--

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Abstract:	The "Melbourne technique" was described in 2008 as a novel method for complete correction of scaphocephaly. Since 2015, it has become our operation of choice for children with sagittal synostosis who are too old at presentation for minimally invasive techniques. Our modifications were 2 position (initially supine then prone) technique and undertaking a formal fronto-orbital remodeling to correct forehead contour. Retrospective chart review was used to record demographics, blood transfusion frequency and volumes, operating time, length of stay, clinical outcome, and complications. 11 underwent modified Melbourne procedure (MMP) between July 2015 and March 2017. 9 of 11 were male. All had a diagnosis of non-syndromic sagittal synostosis. Mean age at surgery was 29 months. Mean surgical time was 6 hours. All patients required blood transfusion with a mean volume transfused of 29 mL/kg (range 13-83 mL/kg). For those 5 patients where pre- and post-operative measurements were available, there was an increase in mean cephalic index (CI) from 0.64 to 0.75. All post-operative patients had a CI of over 0.70. 3D shape analysis indicated head shape change addressing all phenotypic aspects of scaphocephaly. In the 5 patients in which

analysis could be undertaken, the mean intracranial volume increased from 1481 cm³ pre-operatively to 1671 cm³ post-operatively, a mean increase in intracranial volume of 14%. The post-operative intracranial volume was higher than pre-operative in all 5 patients. There were 4 minor and no major complications. MMP is safe and effective for the treatment of severe scaphocephaly in sagittal synostosis.

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Editorial Board
Journal of Craniofacial Surgery

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Dear Editorial Board,

Re: Submission of “Results following adoption of a modified “Melbourne technique” of total scaphocephaly correction at Great Ormond Street Hospital” by Sharma JD et al.

We are pleased to submit the above manuscript for consideration for publication in JCFS.

Whilst isolated sagittal synostosis (ISS) being the most common form of craniosynostosis, a diverse range of surgical techniques have been described, with no consensus on which is best. In 2008, the Melbourne Craniofacial Unit published a novel technique of total calvarial remodelling aiming at complete correction of the phenotype. This study is the first from a different craniofacial unit to report results using this technique – in 11 children.

As the original study, we have found the surgical technique to be safe and effective and report these outcomes, in the form of perioperative data and cranial index. In addition, we have used 3D modelling software to show that the technique reliably increases intracranial volume, and to graphically illustrate the shape change achieved. As we use a minimally invasive technique (springs) for infants with ISS, we have modified the originally described procedure to better serve the older children (average age 2-3 years) in which we perform total calvarial remodelling - and describe and illustrate these modifications in this paper. We believe our findings will be of great interest to all craniofacial surgeons and neurosurgeons who manage children with ISS.

We thank you in advance for your time in considering our submission.

Yours sincerely,



Greg James (on behalf of the authors)

Results following adoption of a modified “Melbourne technique” of total scaphocephaly correction at Great Ormond Street Hospital

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Short title: Modified “Melbourne technique” for scaphocephaly

Statement: No financial or material support were received for the work undertaken in this paper. None of the authors have any conflicts of interest to declare. Written parental consent was obtained for publication of clinical photographs.

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Abstract

The “Melbourne technique” was described in 2008 as a novel method for complete correction of scaphocephaly. Since 2015, it has become our operation of choice for children with sagittal synostosis who are too old at presentation for minimally invasive techniques. Our modifications were 2 position (initially supine then prone) technique and undertaking a formal fronto-orbital remodeling to correct forehead contour. Retrospective chart review was used to record demographics, blood transfusion frequency and volumes, operating time, length of stay, clinical outcome, and complications. 11 underwent modified Melbourne procedure (MMP) between July 2015 and March 2017. 9 of 11 were male. All had a diagnosis of non-syndromic sagittal synostosis. Mean age at surgery was 29 months. Mean surgical time was 6 hours. All patients required blood transfusion with a mean volume transfused of 29 mL/kg (range 13-83 mL/kg). For those 5 patients where pre- and post-operative measurements were available, there was an increase in mean cephalic index (CI) from 0.64 to 0.75. All post-operative patients had a CI of over 0.70. 3D shape analysis indicated head shape change addressing all phenotypic aspects of scaphocephaly. In the 5 patients in which analysis could be undertaken, the mean intracranial volume increased from 1481 cm³ pre-operatively to 1671 cm³ post-operatively, a mean increase in intracranial volume of 14%. The post-operative intracranial volume was higher than pre-operative in all 5 patients. There were 4 minor and no major complications. MMP is safe and effective for the treatment of severe scaphocephaly in sagittal synostosis.

Introduction

Craniosynostosis, defined as the premature fusion of one or more of the skull vault sutures, affects around 1 in 2000 live births, with scaphocephaly due to isolated sagittal synostosis making up approximately 50% of all presentations [1-3]. Treatment is offered primarily to correct head shape for appearance reasons, while opinions regarding the effect of surgery on intracranial pressure and neurological development remain controversial[4-8]. Although there has been recent interest in the use of molding helmet orthoses without operation[9], mainstream treatment remains limited to surgery, and many different techniques have been described, with no clear evidence of superiority of any one method over another[10].

At Great Ormond Street Hospital (GOSH) Craniofacial Unit, the technique of choice in small infants (<6 months of age) with scaphocephaly is spring-assisted cranioplasty, a minimally invasive molding technique[11]. This operation has supplanted our use of vault remodeling techniques such as the “pi” procedure[12] due to reduced operative time, transfusion rates and length of stay[11]. However, it is ineffective in older children whose skulls have lost the visco-elastic properties favourable for molding. For children too old for springs, we have traditionally undertaken a total calvarial remodeling operation involving biparietal widening, barrel staving and a degree of AP shortening. Whilst this operation corrected scaphocephaly and temporal indrawing, it had little effect

on the occipital “bullet” and low posterior vertex which form part of the complex 3-dimensional deformity of scaphocephaly[13].

The “Melbourne technique”, developed by the Melbourne Craniofacial Unit, is a radical method of total cranial vault remodeling which addresses all the phenotypic aspects of scaphocephalic deformity[14]. In addition to frontal and occipital reconstruction, it involves the removal of a strip of bone immediately posterior to the coronal strip (the “A Band”), which is then rotated and replaced at 90 degrees posteriorly to elevate and re-contour the new occiput. This unique step serves to correct the low-lying posterior vertex and creates increased intracranial volume under the newly raised posterior vertex. Total cranial vault remodeling using the Melbourne technique has the advantage of creating a more regular head shape without decreasing calvarial volume. This is achieved with the increased volume under the posterior vertex compensating for the volume lost by the anterior-posterior squeeze. Postoperative helmeting is not required and occipital bulleting, vertex position, and frontal bossing can be corrected in a single procedure.

Due to this potential benefits in both aesthetic outcome and intracranial volume, we adopted the “Melbourne technique”, with some modifications (see Methods), as our standard operation for children with scaphocephaly too old for spring-assisted cranioplasty in 2015. We report our results, the first series since the original paper to do so.

Materials and methods

Patient selection

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[15] supplemented in some cases by radiologic imaging (plain radiograph or three-dimensional computed tomographic 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 aged 6 months or younger, conservative management or spring-assisted cranioplasty; for older children, conservative management or total calvarial vault remodeling using the modified Melbourne procedure (MMP). The prospective advantages and disadvantages of each management strategy are explained and the final decision is made by the family. Patients with multisutural or syndromic craniosynostosis, and those whose parents declined surgical intervention, were excluded from the analysis.

Operative technique

Total calvarial remodeling using the technique described the Melbourne Craniofacial Unit[14] was used, with significant modifications as follows.

We undertake the operation as a two position procedure (under the same general anaesthetic), beginning surgery with the patient supine for fronto-orbital remodeling and removal of the A-band. We then undertake temporary closure with nylon sutures, cover the incision with a sterile occlusive dressing, reposition the patient to a prone position, and re-prepare and drape the field, before performing the parietal and occipital craniotomies using the standard Melbourne method. Undertaking the second stage in the prone position allows the osteotomies around the torcula and major venous sinuses to be carried out with greater visualisation and access. It also permits construction of the neo-occiput (using the A and B flaps) in alignment with the neck. Fixation is with fine steel wires. The C-flaps are inset per the original description and fixed with wires. Bone gaps are filled with ‘salami’, a morsellised bone paste[16].

We undertake a more comprehensive fronto-orbital remodeling, with resection of the supraorbital ridges and temporal squames, dividing the frontal (F) bone block into 2 halves and using the flatter, more posterior part of the bone to reconstruct the forehead and eliminate the frontal bossing (Figure 1). This is in contrast to the Melbourne group’s technique of barrel-staving where the frontal (F) bone flap is left as one piece.

Data Acquisition

To evaluate our outcomes, demographic data, transfusion volumes, and adverse events were obtained from our prospectively maintained craniofacial database. This information was supplemented by a retrospective electronic and paper chart review to gather specific

information on operating time, length of stay, as well as intra- and post-operative complications.

Cephalic index (CI) was calculated from pre-operative CT scans. When post-operative CT scans were available, post-operative CIs were calculated. When both pre- and post-operative CT scans were available for analysis, we performed volumetric analyses to calculate the change in cranial volume. We also performed three-dimensional handheld optical scanning to quantify changes in head shape in a subset of patients.

CT volumetric analysis

For volumetric analysis of CT, semi-automatic segmentation using the commercial software Simpleware Scan IP (Simpleware Ltd., Exeter, UK), running on 64-bit Operating System with Intel Xeon CPU E3-1270 and Windows 7 Enterprise, Service Pack 1 (Microsoft Corporation) was used. DICOM images were loaded into Simpleware and subsequently rotated and cropped to include foramen magnum to vertex. The image threshold was set at -55HU to 117HU for all scans, similar to others published in the literature[17]; these parameters were found visually to provide the most useful soft tissue range for the first mask, highlighting the region of interest to be created. Intracranial contents were separated from the surrounding tissues using a region growing operation (known as ‘flood fill’ in Simpleware); here the software fills in connected regions of the mask using a seed point and the given threshold. After this, the spill of the mask from the skull base foramina was assessed, and initially corrected through a series of open and

close morphological operations. Any remaining spill that could not be solved using the morphological operations was removed manually by closing the remaining cranial defects. This produced a final mask that best filled the intracranial cavity across axial, sagittal and coronal views. The volume of this mask was calculated, based on mask statistics in Simpleware, using the voxel information within the mask.

3D optical scanning: image acquisition and processing

3D scans of 2 patients with sagittal synostosis were acquired pre- and post-TCR in theatre using a structured light handheld scanner (M4D Scanner, Rodin4D, Pessac, France) connected to a laptop with VXelements software (Creaform, Levis, Quebec, Canada). Since structured light handheld cameras have difficulties capturing hair, tight white nylon stockings (Beagle Orthopaedic, UK) were used to cover patient hair.

Scans were post-processed following the steps described by our group previously[18]. Briefly, scans were exported as 3D computational surface meshes in stereolithography (STL) format and imported to MeshMixer software (Autodesk Inc., Toronto, ON, Canada) to clean artefacts. Multiple incomplete scans of the same patient were merged together using 3-matic (Materialise, Leuven, Belgium) to create a full 3D model. The models were cut by a plane defined by the nasion and both tragion using Meshmixer to focus on the region which is remodelled during TCR (i.e. calvarium). Measurements were taken on the cleaned and cut pre- and post-operative scans of each patient with Meshmixer. Head length and width were measured to calculate the CI. Head volume was understood as the volume under the mesh surface capped by the cutting plane.

Results

Demographics

11 consecutive patients were identified who underwent MMP for scaphocephaly between July 2015 and March 2017. All patients had non-syndromic isolated sagittal synostosis. There were 9 males and 2 females. Mean age at surgery was 29 months (range 18-43 months). Mean weight at surgery was 13 kg (range 10-17 kg).

Operative and hospital data

The mean operative time for MMP, including induction of anaesthesia, was 6.5 hours (range 3.5-8 hours). Mean pre-operative haemoglobin (in g/mL) was 119 (range 109-131 g/mL). All patients required blood transfusion intra-operatively, with a mean volume transfused (packed red blood cells only) of 29 mL/kg (range 13-83 mL/kg). Mean volume of packed red cells transfused was 433 mL (range 150-1162 mL). 10 patients had 1 donor exposure, with 1 patient having 2 donor exposures. Mean day 1 post-operative haemoglobin was 111 g/mL (range 95-148 g/mL), with 10 of 11 patients having a reduced haemoglobin post-operatively (mean decrease of 11 g/mL). The 1 patient with a higher post-operative haemoglobin received the relatively large (83 mL/kg) intra-operative transfusion and 2 donor exposures. They did not experience any clinical ill-effects from the relatively larger transfusion. The mean length of hospital stay was 5 days (range 4-6 days). No patient stayed in hospital longer than 6 days.

Cephalic index

Pre-operative CI was available for all 11 patients. All had significant scaphocephaly with a mean CI of 0.64 (range 0.57-0.69). In addition to low CI, all had typical phenotypic features of isolated sagittal synostosis with frontal bossing, parietal narrowing, occipital “bullet” and anterior displacement of the vertex (Figure 2). Post-operative CI was available for 5 patients, 3 measured on CT and 2 measured using 3D photography. Post-operatively, CI was increased in all 5 patients, with a mean post-operative CI of 0.75 (range 0.74-0.77; Figure 3).

3D imaging and volumetric analysis

Pre- and post-operative three-dimensional optical scanning was performed on 2 patients in the series. From this data, we were able to construct three-dimensional distance maps demonstrating head shape change following MMP (Figure 4). These maps show the reduction of frontal bossing, correction of the anteriorly displaced vertex, increase in biparietal diameter, and decrease in occipital “bullet” after surgery.

Pre- and post-operative calvarial volumes could be calculated in 5 patients. All patients had an increased calvarial volume following surgery. The mean calvarial volume increased from 1481 cm³ pre-operatively (range 1270-1891 cm³) to 1671 cm³ post-operatively (range 1337-20082 cm³), representing a mean increase in intracranial volume

of 14% (Figure 5). In these 5 patients, all had an increase in intracranial volume post-operatively (range of increase 117 – 274 cm³), with no patient having a reduced volume.

Complications

There were 4 complications in our group of 11 patients, all of them minor using the standard UK craniofacial classification (class II or III; Table 1)[11]. Class II complications were as follows ($n = 2$): re-admission 13 days after surgery for a viral respiratory illness and anaemia (haemoglobin 76 g/L) requiring a blood transfusion; and CSF leak from incision (post-operative day 1) requiring a stitch placed on the ward. Class III complications were as follows ($n = 2$): wire protrusion requiring a second surgery one month post-operatively; and minor dehiscence of bicoronal incision following minor injury requiring re-suture in the operating room. There were no neurological complications (grade IV) and no mortality (grade V).

Follow-up

Mean follow up was 511 days (range 345-725 days). All patients had significant subjective improvement in head shape as demonstrated by clinical photographs and feedback from families (Figure 6). As yet, no patients or families have requested any further surgery for appearance reasons, and no child has developed clinical, radiological or ophthalmological evidence of raised intracranial pressure. At GOSH, all single-suture

children are followed until at least age 12, allowing detection and reporting of late complications.

Discussion

The original report of the Melbourne Craniofacial Unit of their novel technique for correction of scaphocephaly demonstrated that, in their hands, the method was safe and efficacious[14]. The current study is the first from another centre to report experience with the technique.

Our experience in these 11 cases broadly reflects that of the original series: that the “Melbourne technique” is a safe and effective procedure for the correction of scaphocephaly. Whilst operative times (mean of 6 hours) were relatively long, especially in comparison to our minimally invasive spring-assisted cranioplasty used in infants[11], they are not particularly excessive when compared to operative times in cranial vault remodeling surgeries in general[19]. No serious intra- or post-operative complications were encountered, with no neurological morbidity and no mortality. 4 patients had minor complications, with 2 requiring visits to the operating room, 1 for removal of a protruding wire and 1 for partial resuturing of the bicoronal incision. For such a major vault remodeling procedure we find this an acceptable complication profile. In terms of effectiveness, there was an increase in CI seen in all patients into the “normal” range following MMP (usually quoted as >0.70)[20]. Increasingly, change in CI alone is seen as an over-simplification of the deformity and correction of scaphocephaly[21]. Our 3D imaging studies comparing shape pre- and post-MMP indicate that, as well as an

increased CI there is a significant improvement in frontal bossing, occipital deformity and vertex position (Figure 4).

Effect on intracranial volume

One of the concerns regarding all techniques for surgical correction of sagittal synostosis is the potential for post-operative deleterious effects on skull growth and intracranial volume. In fact, several studies have suggested that traditional vault remodeling techniques may reduce intracranial volume[12, 22-24]. As restriction of cranial volume appears to be a risk factor for raised intracranial pressure[6, 25, 26], it seems logical that techniques that increase (or at least stabilize) intracranial volume are preferable to those that do not. It was reassuring to note that in the 5 children in the series for which we were able to calculate pre- and post-operative intracranial volumes all showed an increase – with a mean of 14% across the series. We have yet to encounter a case of raised intracranial pressure in this series, but longer follow-up is required to establish whether or not this increase in intracranial volume in MMP correlates with any protective clinical effect on reducing intracranial hypertension.

Modifications from originally described technique

We have made some modifications to the originally described Melbourne technique for our practice. We believe that these technical modifications have been predominantly dictated by the difference in age in our cohort – our mean age at surgery was 29 months,

in comparison to 7.5 months in the original Melbourne paper[14]. Older children have comparatively smaller heads and less flexible necks, as well as thicker and less malleable bone[27]. We therefore felt that the initial description of performing the entire surgery in the supine position would be suboptimal due to the relative restriction of visualization of the occipital osteotomies, particularly as these bone cuts are over the major venous sinuses and torcula. This led to our modification of performing the surgery in 2 stages under the same general anaesthetic, with a change of position from supine to prone in between. There are theoretical risks of intra-operative change of position including anaesthetic issues and de-sterilization of the operative field, but we did not see these in our series. Our experience suggests that communication between the surgical, anaesthetic and nursing team is key to ensure this changeover is smooth and safe. Our mean surgical time was around 1 hour longer than that described in the original study[14], but our transfusion volumes were slightly lower (433 ml/case vs 460 ml/case, in larger children), suggesting that the re-opening for the second stage does not lead to greater haemorrhage. We did not see any infections in our cohort, which again supports the contention that careful protection of the wound and re-draping during the change of position is sufficient to prevent de-sterilisation of the field.

The other major modification is how the bossing of the frontal (F) flap is dealt with. In the original paper, simple barrel staving is used to recontour the forehead[14]. We initially attempted to replicate this method, but were unable to create an acceptable on-table contour. Again, we contend that this is due to the older age with thicker, less flexible bone. Therefore, in all of our cases we undertook our standard fronto-orbital

remodeling technique, dividing the frontal (F) bone block into 2 halves, rotating them and fixing them in a widened position to correct both the frontal bossing and the temporal in-drawing (Figure 7). One potential issue in undertaking a complex vault remodeling with volume expansion and multiple osteotomies in older children is the potential for multiple bony “gaps”. This is less of an issue in infants whose dura has much greater osteogenic potential, with older age at craniofacial surgery being a risk factor for gaps[28]. We address this by using our standard technique of “salami” morcellised bone paste[16] to fill any gaps in the reconstruction. As yet, there have not been any issues with bone defects in this series, but longer follow-up will need to be reported to ensure this is a sustained effect. Our modifications appear to negate the possible unfavorable aspects of operating in our older age group, and suggest that the MMP is an appropriate and effective operation for correction of scaphocephaly in this cohort.

Conclusions

The Melbourne Craniofacial Unit published a novel technique for surgical correction of scaphocephaly[14] which addresses phenotypic aspects such as vertex position and occipital deformity that are traditionally difficult to treat. Results from our series, the first from a different centre, support the contention that it is an effective and safe operation. We suggest that our modifications have successfully adapted the technique to an older, non-infant cohort of patients.

References

1. Kweldam CF, van der Vlugt JJ, van der Meulen JJNM (2011) The incidence of craniosynostosis in the Netherlands, 1997-2007. *British Journal of Plastic Surgery* 64:583–588. doi: 10.1016/j.bjps.2010.08.026
2. Lee HQ, Hutson JM, Wray AC, et al (2012) Changing epidemiology of nonsyndromic craniosynostosis and revisiting the risk factors. *Journal of Craniofacial Surgery* 23:1245–1251.
3. Cornelissen M, Ottelander BD, Rizopoulos D, et al (2016) Increase of prevalence of craniosynostosis. *Journal of Cranio-Maxillofacial Surgery* 44:1273–1279. doi: 10.1016/j.jcms.2016.07.007
4. Hayward R, Britto J, Dunaway D, Jeelani O (2016) Connecting raised intracranial pressure and cognitive delay in craniosynostosis: many assumptions, little evidence. *Journal of Neurosurgery: Pediatrics* 18:242–250. doi: 10.3171/2015.6.PEDS15144
5. Hayward R, Britto JA, Dunaway D, et al (2015) Raised intracranial pressure and nonsyndromic sagittal craniosynostosis. *Journal of Neurosurgery: Pediatrics* 16:346–348. doi: 10.3171/2014.11.PEDS14625
6. Wall SA, Thomas GPL, Johnson D, et al (2014) The preoperative incidence of raised intracranial pressure in nonsyndromic sagittal craniosynostosis is underestimated in the literature. *Journal of Neurosurgery: Pediatrics* 14:674–681.

doi: 10.3171/2014.8.PEDS1425

7. Patel A, Yang JF, Hashim PW, et al (2014) The impact of age at surgery on long-term neuropsychological outcomes in sagittal craniosynostosis. *Plastic and Reconstructive Surgery* 134:608e–17e.
8. Bellew M, Chumas P (2015) Long-term developmental follow-up in children with nonsyndromic craniosynostosis. *Journal of Neurosurgery: Pediatrics* 16:445–451. doi: 10.3171/2015.3.PEDS14567
9. Sood S, Rozzelle A, Shaqiri B, et al (2011) Effect of molding helmet on head shape in nonsurgically treated sagittal craniosynostosis. *Journal of Neurosurgery: Pediatrics* 7:627–632. doi: 10.3171/2011.4.PEDS116
10. Doumit GD, Papay FA, Moores N, Zins JE (2014) Management of Sagittal Synostosis. *Journal of Craniofacial Surgery* 25:1260–1265. doi: 10.1097/SCS.0b013e3182a24635
11. Rodgers W, Glass GE, Schievano S, et al (2017) Spring-Assisted Cranioplasty for the Correction of Nonsyndromic Scaphocephaly: A Quantitative Analysis of 100 Consecutive Cases. *Plastic and Reconstructive Surgery* 140:125–134. doi: 10.1097/PRS.0000000000003465
12. Boop FA, Chaddock WM, Shewmake K, Teo C (1996) Outcome analysis of 85 patients undergoing the pi procedure for correction of sagittal synostosis. *Journal of Neurosurgery* 85:50–55. doi: 10.3171/jns.1996.85.1.0050

13. Bendon CL, Johnson HP, Judge AD, et al (2014) The aesthetic outcome of surgical correction for sagittal synostosis can be reliably scored by a novel method of preoperative and postoperative visual assessment. *Plastic and Reconstructive Surgery* 134:775e–786e.
14. Greensmith AL, Holmes AD, Lo P, et al (2008) Complete correction of severe scaphocephaly: the Melbourne method of total vault remodeling. *Plastic and Reconstructive Surgery* 121:1300–1310. doi: 10.1097/01.prs.0000304592.56498.d6
15. Cerovac S, Neil-Dwyer JG, Rich P, et al (2009) Are routine preoperative CT scans necessary in the management of single suture craniosynostosis? *British Journal of Neurosurgery* 16:348–354. doi: 10.1080/0268869021000007560
16. Rashid A, Marucci DD, Dunaway DJ, Hayward RD (2008) Bone “salami”: morcellised bone and fibrin glue for filling extensive cranial defects in craniofacial surgery. *British Journal of Plastic Surgery* 61:993–996. doi: 10.1016/j.bjps.2007.10.056
17. Leikola J, Koljonen V, Heliövaara A, et al (2014) Cephalic index correlates poorly with intracranial volume in non-syndromic scaphocephalic patients. *Childs Nerv Syst* 30:2097–2102. doi: 10.1007/s00381-014-2456-x
18. Tenhagen M, Bruse JL, Rodriguez-Florez N, et al (2016) Three-Dimensional Handheld Scanning to Quantify Head-Shape Changes in Spring-Assisted Surgery for Sagittal Craniosynostosis. *Journal of Craniofacial Surgery* 27:2117–2123.

19. Stricker PA, Goobie SM, Cladis FP, et al (2017) Perioperative Outcomes and Management in Pediatric Complex Cranial Vault Reconstruction. *Anesthesiology* 126:276–287. doi: 10.1097/ALN.0000000000001481>
20. van Veelen M-LC, Eelkman Rooda OHJ, de Jong T, et al (2013) Results of early surgery for sagittal suture synostosis: long-term follow-up and the occurrence of raised intracranial pressure. *Childs Nerv Syst* 29:997–1005. doi: 10.1007/s00381-013-2024-9
21. Fearon JA, Ditthakasem K, Herbert M, Kolar J (2017) An Appraisal of the Cephalic Index in Sagittal Craniosynostosis, and the Unseen Third Dimension. *Plastic and Reconstructive Surgery* 140:138–145.
22. Arab K, Fischer S, Bahtti-Softeland M, et al (2016) Comparison Between Two Different Isolated Craniosynostosis Techniques: Does It Affect Cranial Bone Growth? *Journal of Craniofacial Surgery* 27:e454–7. doi: 10.1097/SCS.0000000000002769
23. Fischer S, Maltese G, Tarnow P, et al (2016) Comparison of Intracranial Volume and Cephalic Index After Correction of Sagittal Synostosis With Spring-assisted Surgery or Pi-plasty. *Journal of Craniofacial Surgery* 27:410–413.
24. Heller JB, Heller MM, Knoll B, et al (2008) Intracranial volume and cephalic index outcomes for total calvarial reconstruction among nonsyndromic sagittal synostosis patients. *Plastic and Reconstructive Surgery* 121:187–195.
25. Sood S, Marupudi N, Haridas A, Ham SD (2015) Intracranial pressure and sagittal

craniosynostosis. *Journal of Neurosurgery: Pediatrics* 16:351–352. doi: 10.3171/2015.1.PEDS14705

26. Florisson JMG, van Veelen M-LC, Bannink N, et al (2010) Papilledema in isolated single-suture craniosynostosis: prevalence and predictive factors. *Journal of Craniofacial Surgery* 21:20–24. doi: 10.1097/SCS.0b013e3181c3465e
27. Delye H, Clijmans T, Mommaerts MY, et al (2015) Creating a normative database of age-specific 3D geometrical data, bone density, and bone thickness of the developing skull: a pilot study. *Journal of Neurosurgery: Pediatrics* 16:687–702. doi: 10.3171/2015.4.PEDS1493
28. Noordzij N, Brouwer R, van der Horst C (2016) Incomplete Reossification After Craniosynostosis Surgery. *Journal of Craniofacial Surgery* 27:e105–8.

Figure Legends

Figure 1. Diagram illustrating our modification of the Melbourne technique. The surgery is conducted in 2 stages under the same general anaesthetic. Left: the child is placed in the supine position to allow craniotomies of the “F” frontal flap and the “A” band. The F flap is reconstructed using our standard fronto-orbital remodeling technique. The A band is kept sterile and used in the second stage to form the new occiput. Right: the incision is temporarily closed and kept sterile and the child turned prone. This allows excellent visualization during the “B” and “C” flap osteotomies.

Figure 2. Pre-operative CT 3D reconstruction from typical case. All patients in our series had a cephalic index of below 0.69. Note the complex nature of scaphocephalic deformity, with frontal bossing, temporal in-drawing (“pinching”), anteriorly placed vertex and an occipital “bullet” deformity.

Figure 3. Box-and-whisker plot to show change between pre- and post-operative cephalic index (CI) following modified Melbourne procedure. There was an increase in mean CI from 0.64 to 0.75. All post-operative patients had an increased CI, and all had a post-operative CI of over 0.70.

Figure 4. Distance change “heat” maps demonstrating post-operative shape change following modified Melbourne procedure. “Hot” colours demonstrate an increased external projection from pre-op, whilst “cold” colours represent retrusion towards the midpoint of the skull. In these 2 examples, both patients had relative retrusion (blue) of their previously bossed foreheads and occiputs, whilst the parietal bones have been widened (red). This is consistent with the intended correction of the complex deformity of scaphocephaly.

Figure 5. Change in intracranial volume (ICV) following modified Melbourne procedure. Pre-operative (open) and post-operative (shaded) ICV in the 5 patients where volumes could be calculated. There was a mean increase of 14% of ICV, and no patient had a reduced ICV following MMP.

Figure 6. Clinical photographs illustrating results of modified Melbourne procedure. 2 patients are illustrated. The global effect of correction on all phenotypic aspects of scaphocephaly, including forehead contour, vertex position and occipital shape, as well as improvement in CI, can be appreciated.

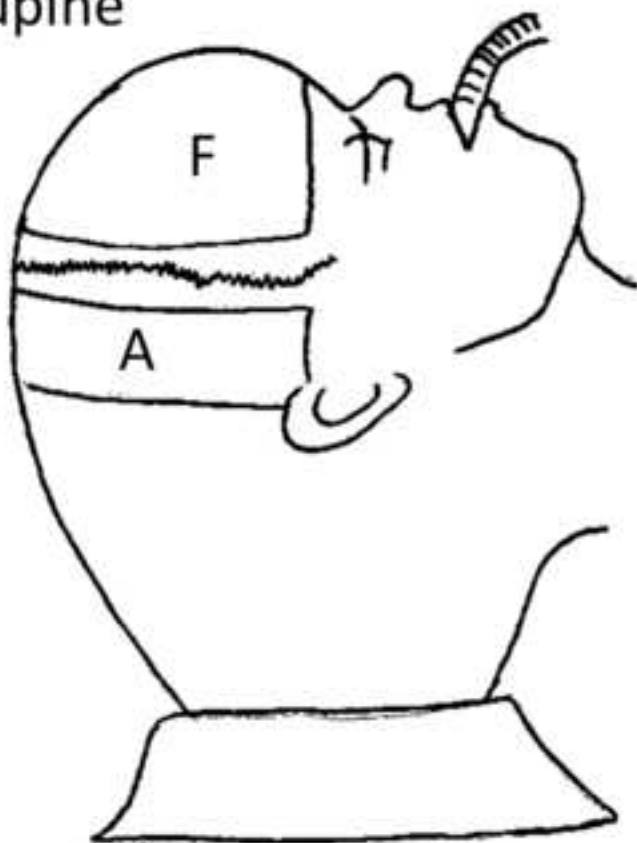
Figure 7. Pre and post-operative CT scan demonstrating modified Melbourne procedure. Our modifications, including the use of a more formal fronto-orbital remodeling technique rather than simple barrel-staving of the “F” flap can be appreciated.

Note how the majority of the bony gaps have re-ossified by the time of this CT scan (6 months post-op). We use morcellised bone paste (“salami”) to fill all osseous defects at the time of surgery.

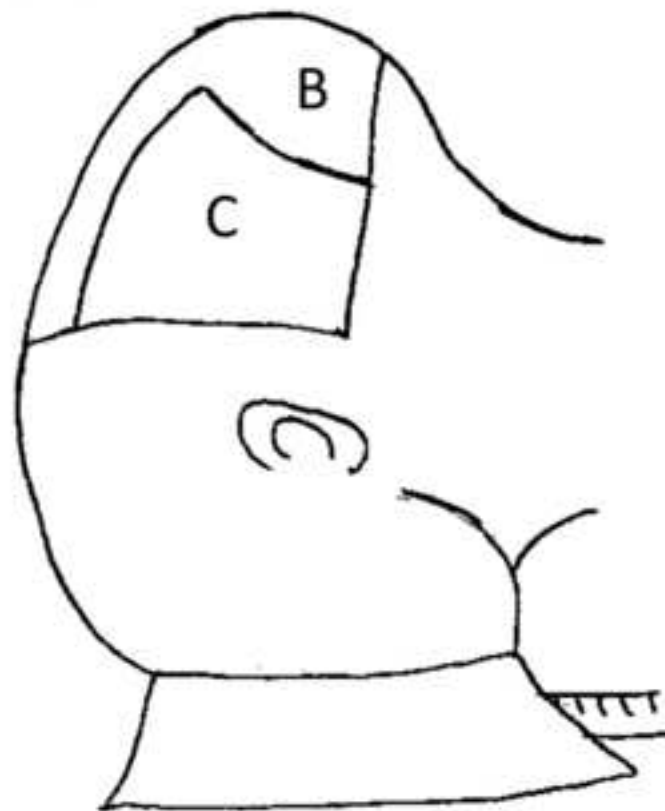
Table Legends

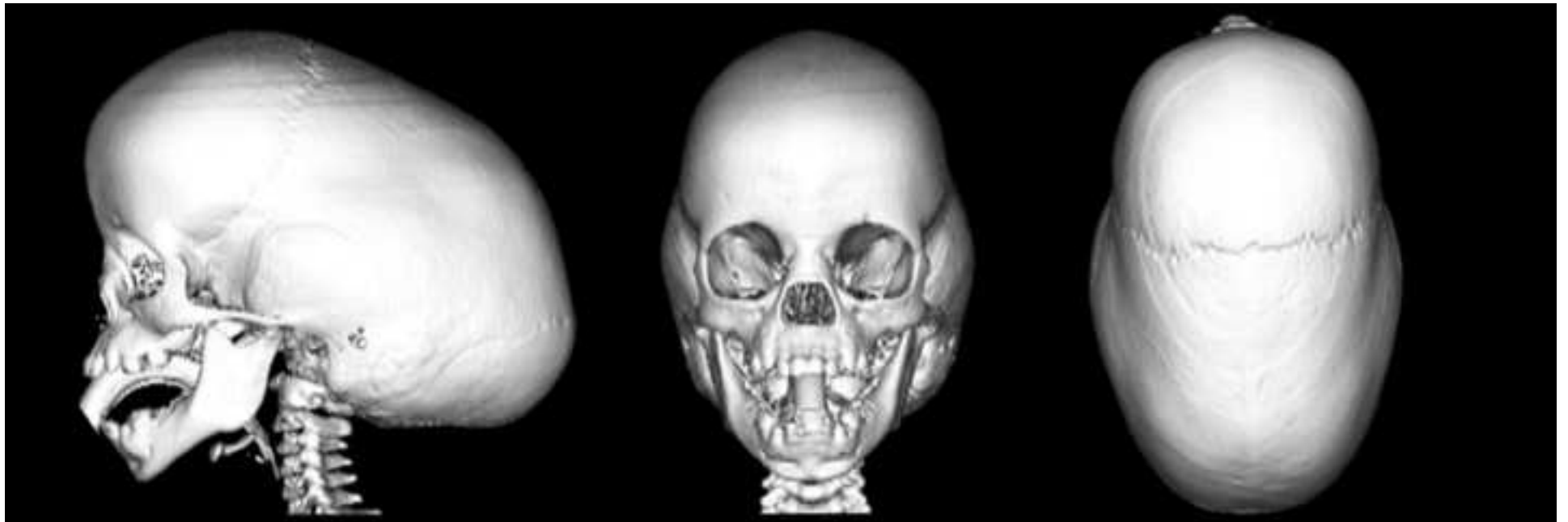
Table 1. Complications arising from modified Melbourne procedure in the current series. We use the standard UK craniofacial surgery complication grading developed by the Oxford Craniofacial Unit.

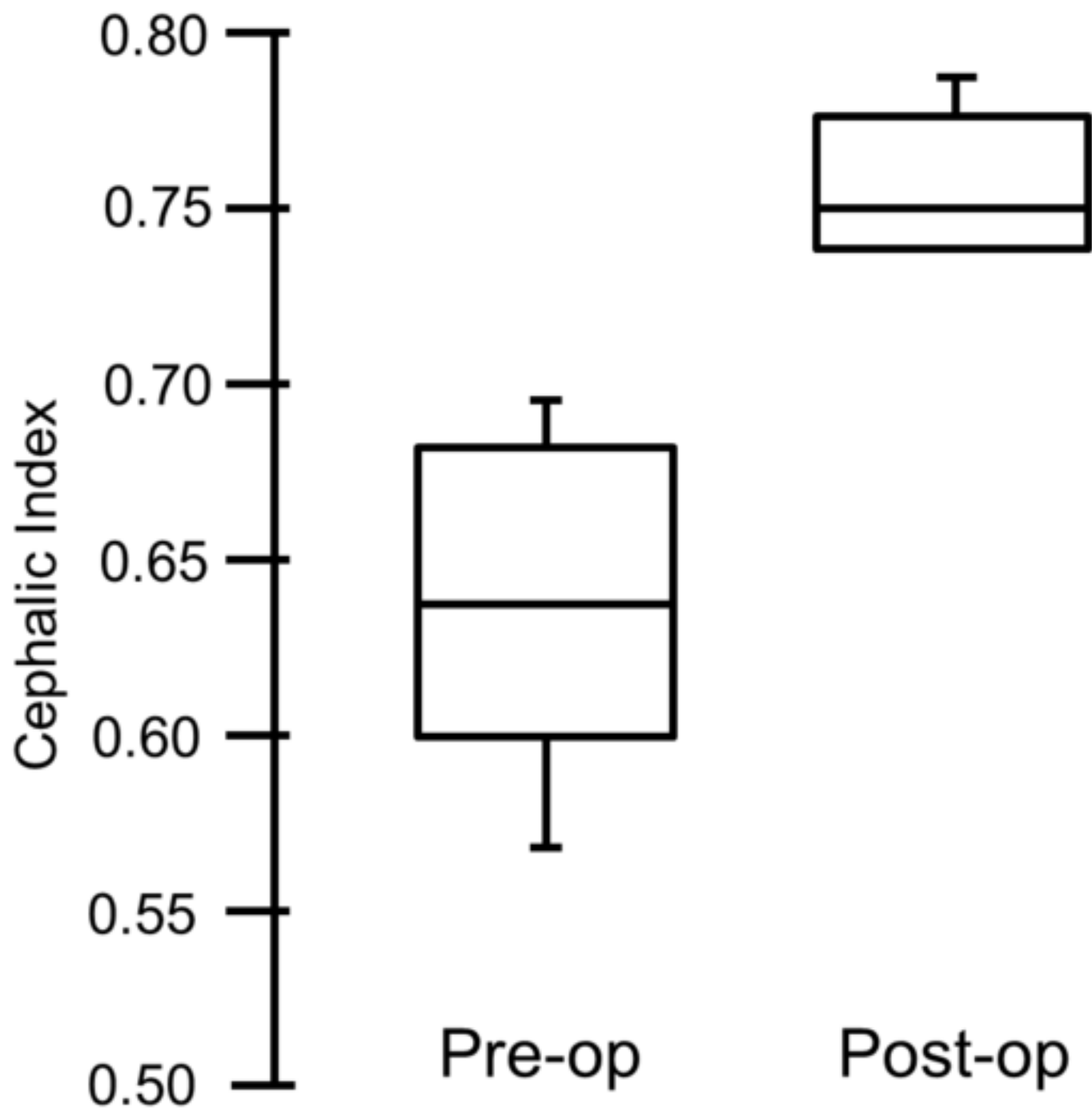
Supine

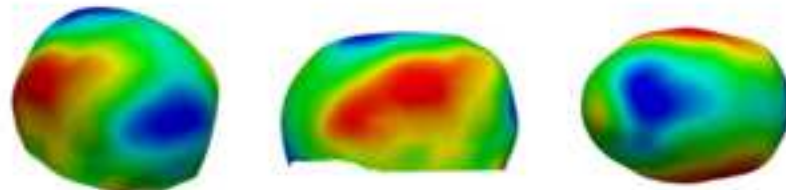
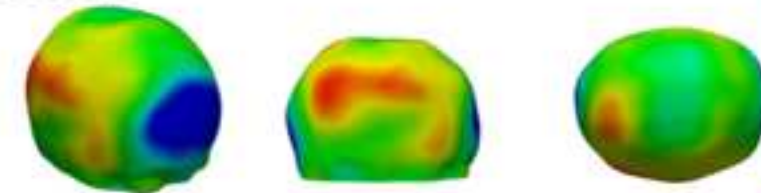


Prone



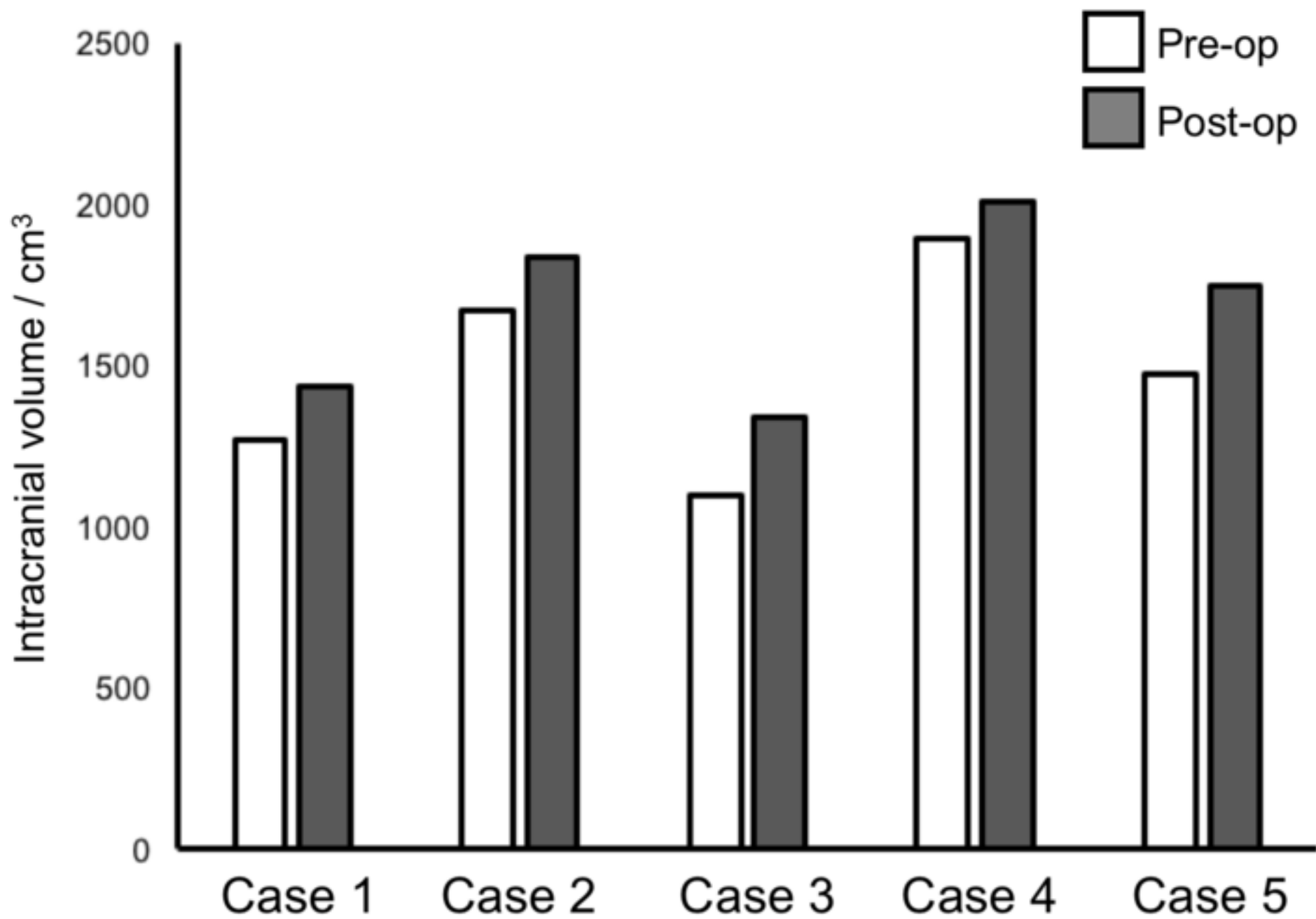




Pre-op**Post-op****Pre-op****Post-op****Pre-op****Post-op****Pre-op****Post-op**

Distance (mm)





Pre-op



Post-op



Pre-op



Post-op



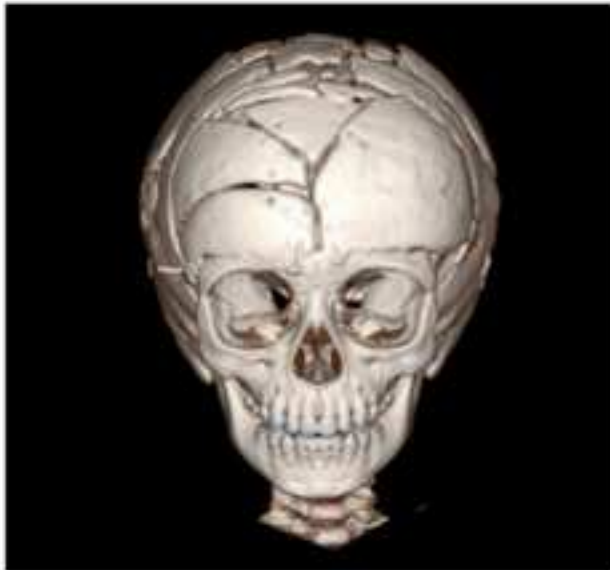
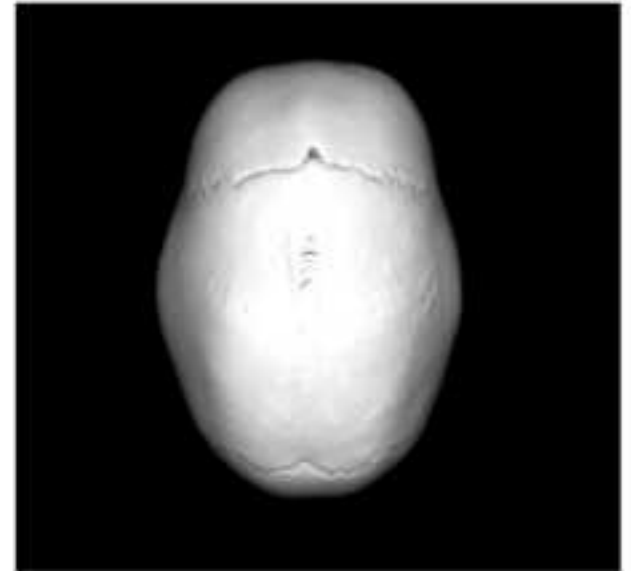
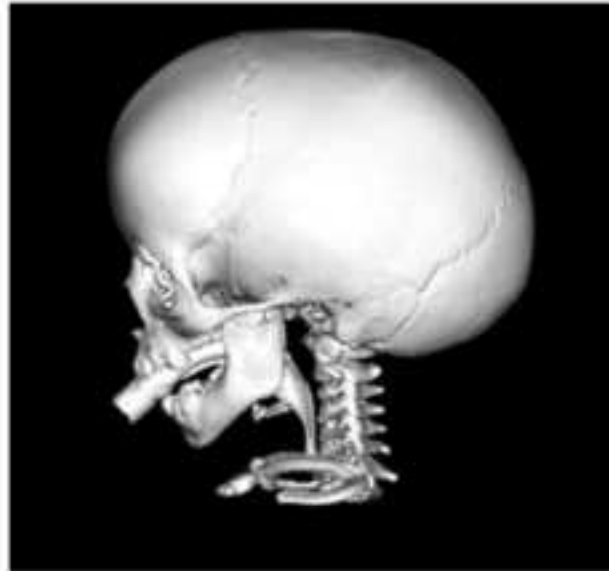


Table 1

Type	Complication	Number encountered
I	No delay in discharge, re-operation or long-term sequelae	0
II	Delay in discharge but no re-operation required	2
III	Re-operation but no long-term sequelae	2
IV	Long-term deficit or neurological impairment or permanent disability	0
V	Mortality	0

RECEIVED

03 AUG 2017

GREAT ORMOND STREET HOSPITAL FOR CHILDREN NHS TRUST CONSENT TO PHOTOGRAPHY OR VIDEO RECORDING

Great Ormond Street Hospital for Children NHS Trust has adopted a policy in line with the Data Protection Act, which gives you the right to control the future use of photographs or video recordings taken of you during the course of your treatment.

***a Referral to Medical Illustration:**

I wish to refer you to the Medical Illustration Department for medical photographs to be taken. These photographs will be part of your medical records and may be used for teaching of medical, paramedical and nursing staff as well as medical students, or for specific other use as detailed below.

***b Medical Photography in the Trust by other staff:**

LESITH DASANAYAKE
** I CHAMALI M. BASTIANGE* confirm that I have registered with Medical Illustration that the photography and the storage of the resulting images will take place in line with the Trust's Policy for making and using illustrative clinical records of patients, and I will take the appropriate photographs in a dignified manner, using equipment approved by Medical Illustration.

This consent limits their use to the purposes only specified by you and should it be desired to use your photograph(s) in any other way – for example, in a medical textbook or an on-line teaching resource – the Trust will seek your specific permission to do so.

Clinician's name: *MR H JAMES* (print) Speciality: *CRANIOPACUR*

Clinician's signature: 

CONSENT

In view of the explanation given to me by Prof/Dr/Mr/Miss/Mrs: *JAMES*

- I consent to photographs being taken for my personal medical case-notes.
- I consent to photographs being made available for teaching in the Healthcare context as described above.
- I consent to my photographs being published for the specific purpose described below. This consent does not extend to any further publication(s).

SCIENTIFIC PAPER ABOUT 'MEASURE TECHNIQUE'

Signature of patient: 

* Signature of parent/guardian: *Mrs CHAMALI M. BASTIANGE* * Date: *23/07/2017*

* Relationship to patient: *Mother*

Top copy to be retained in patient's Medical Records
Second copy to be given to the patient
Third copy to be forwarded to Medical Photography

27 JUL 2017

GREAT ORMOND STREET HOSPITAL FOR CHILDREN NHS TRUST CONSENT TO PHOTOGRAPHY OR VIDEO RECORDING

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***a Referral to Medical Illustration:**

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***b Medical Photography in the Trust by other staff:**

* I CLARISSA HINE confirm that I have registered with Medical Illustration that the photography and the storage of the resulting images will take place in line with the Trust's *Policy for making and using illustrative clinical records of patients*, and I will take the appropriate photographs in a dignified manner, using equipment approved by Medical Illustration.

This consent limits their use to the purposes only specified by you and should it be desired to use your photograph(s) in any other way – for example, in a medical textbook or an on-line teaching resource – the Trust will seek your specific permission to do so.

Clinician's name: MR G JAMES (print) Speciality: CRANIOPACUL

Clinician's signature [Signature]

CONSENT

In view of the explanation given to me by Prof/Dr/Mr/Miss/Mrs: G JAMES

- I consent to photographs being taken for my personal medical case-notes.
- I consent to photographs being made available for teaching in the Healthcare context as described above.
- I consent to my photographs being published for the specific purpose described below. This consent does not extend to any further publication(s).

SCIENTIFIC PAPER ABOUT 'METABOLIC TERMINANT'

Signature of patient:

* Signature of parent/guardian: Clarissa Hine Date: 24 July 2017

* Relationship to patient: MOTHER

Top copy to be retained in patient's Medical Records
Second copy to be given to the patient
Third copy to be forwarded to Medical Photography