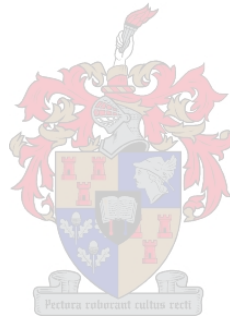


**MODIFIED TOTAL CRANIAL REMODELING TECHNIQUE FOR
SCAPHOCEPHALY REPAIR**

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Declaration

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Abstract

Introduction: Sagittal synostosis or scaphocephaly is the most common isolated single-suture synostosis that accounts for 40% to 60% of all craniosynostosis cases which affects 1 out of 2000 live births. The craniofacial unit at Tygerberg Academic Hospital modified the technique of total vault remodeling by lag screw fixation of onlay bone segments in the temperoparietal region to: a) improve the stability of the reconstruction; b) to increase the biparietal distance; c) to reduce operation time; and lastly d) to avoid secondary procedures for the removal of titanium plates. The aim of this study was to evaluate the surgical outcomes of the modified total cranial vault remodeling procedure for the management of sagittal synostosis.

Method: A retrospective study was employed to investigate the surgical outcomes of the modified total cranial vault remodeling technique for non-syndromic scaphocephaly repair by use of medical records of eight pediatric patients operated over thirty-two months from October 2011 to May 2014. The sample comprised three boys and five girls with an age range of 4 months to 5 years and 7 months. The head circumference was measured pre- and post-operatively and the parents' satisfaction recorded. The surgical duration of the modified procedure and the patients' blood transfusion volume was compared to the unit's traditional approach.

Results: The head circumference of all patients increased on the percentiles of the head circumference-for-age growth chart. Pre-operatively a mean of 47 cm and post-operatively a mean of 50.94 cm were measured. Parents were generally satisfied with the aesthetic outcomes of the surgery. The average volume for intraoperative blood transfusion was 230 ml compared to 763 ml for the conventional method. The average surgical time decreased from 5.5 hours with the conventional method to 3.4 hours with the modified technique.

Conclusion: The modification of the cranial vault remodeling increased the head circumference, yielded good parental satisfaction, decreased the surgery time and intraoperative blood transfusion volume with complications comparable to the traditional method.

Keywords:

Resorbable plates, Single suture craniosynostosis, strip craniectomy, total cranial vault remodeling

Opsomming

Inleiding: Sagittale sinostose of skafosefalie is die mees algemene geïsoleerde enkel-hegting sinostose. Dit is verantwoordelik vir 40-60% van alle kraniosinostose gevalle, en beïnvloed 1 uit 2000 lewende geboortes. Die kraniofasiale eenheid by Tygerberg Akademiese Hospitaal het die tegniek van die totale skedeldak hermodellering verander deur skroeffiksasie van oorvleuel been segmente in die temperoparietal streek gewysig om: a) die stabiliteit van die rekonstruksie te verbeter; b) die biparietale afstand te verhoog ; c) vermindering van operasie tydsduur en laastens, en d) die vermyding van sekondêre prosedures vir die verwydering van titanium plate. Die doel van hierdie studie was om die chirurgiese uitkomst van die gewysigde totale kraniale skedeldak hermodellering prosedure te evalueer in die hantering van sagittale sinostose.

Metode: 'n Retrospektiewe studie is ingestel om die chirurgiese uitkomst van die gewysigde totale kraniale skedeldak hermodellering tegniek vir nie-sindromiese skafosefalie herstel te ondersoek deur die gebruik van mediese rekords van agt pediatriese pasiënte. Hierdie pasiënte is geopereer oor 'n tydperk van 32 maande vanaf Oktober 2011 tot Mei 2014. Die studie bestaan uit drie seuns en vyf meisies met 'n ouderdomsvariasie van 4 -67 maande. Die kopomtrek is pre en postoperatief gemeet en die ouers se tevredenheid met die prosedure was bepaal. Die duur van die chirurgiese gewysigde prosedure en bloedoortapping volume van die pasiënte is vergelyk met die tradisionele benadering.

Resultate: Die kopomtrek van alle pasiënte het vermeerder op die persentiele van die kop omtrek-vir-ouderdom groeikaart. Pre-operatief is 'n gemiddeld van 47 cm en post-operatief 'n gemiddeld van 50.94 cm gemeet. Ouers was oor die algemeen tevrede met die estetiese uitkomst van die operasie. Die gemiddelde volume vir intraoperatiewe bloedoortapping was 230 ml in vergelyking met 763 ml vir die konvensionele metode. Die gemiddelde chirurgiese duur het verminder vanaf 5.5 uur (die tradisionele metode) tot 3,4 uur met die gewysigde tegniek.

Gevolgtrekking: Die wysiging van die kraniale skedeldak hermodellering verhoog die kopomtrek. Dit lewer goeie ouerlike tevredenheid, en dit verminder die operasie tyd en intraoperatiewe bloedoortapping volume met komplikasies vergelykbaar met die tradisionele metode.

1. Introduction

Craniosynostosis is recognized as a group of birth defects that impair the skull structures by the premature fusion of one or more cranial sutures resulting in an abnormal head shape (Aviv, Rodger & Hall, 2002; Rogers, 2011). The early closure of the cranial sutures may result in a variety of functional alterations such as neurocognitive function, breathing, feeding and vision and morphological alterations of the craniofacial development (Aviv, Rodger & Hall, 2002; Ciurea, Toader & Mihalache 2011).

Simple craniosynostosis occurs when only one suture fuses prematurely and complex craniosynostosis occurs when there is premature fusion of multiple sutures (Arseni, Horvath & Ciurea, 1985; Ciurea, Toader & Mihalache, 2011). When children suffering from the complex type display other bodily deformities, they are referred to as syndromic craniosynostosis. Non-syndromic craniosynostosis however predominates and is described as an isolated suture fusion that may result in functional impairment related to local effects of the premature union (Arseni, Horvath & Ciurea, 1985; Ciurea, Toader & Mihalache, 2011).

Sagittal synostosis or scaphocephaly is the most common isolated single-suture synostosis that accounts for 40% to 60% of all craniosynostosis cases (Persing, Jane & Edgerton, 1989) which affects 1 out of 2000 live births (Shillito & Matson, 1968). It has an incidence of 0.4 out of 1000 newborn, with a male and female ratio of 4:1 (Shillito & Matson, 1968; Ciurea, Toader, Mihalache, 2011; Persing, 2008). Scaphocephaly is characterised by the premature closure of the sagittal suture and it is distinctive in its narrow, elongated cranial vault along with reduced biparietal and bitemporal dimensions (Marchac & Arnaud, 1999). The growth of the skull bones does not occur at the sagittal suture, but at the lambdoid and coronal sutures. Since the frontal and occipital bones compensate for the restricted growth of the parietal bones, the elongation of the skull occurs.

Clinically, the head is shortened in the biparietal diameter, elongated in the anterior-posterior diameter and ridging of the sagittal suture may be palpable or visible. Often the fontanelle at the

top of the head is absent. The skull looks elongated in the side view and from the top. It is broader near the forehead and progresses narrower towards the back. In the frontal view of the infant's face, the forehead seems prominent and the sides of the skull narrow. Classically the cephalic index (bitemporal distance divided by anterior postal distance) is from 71 to 76 (Nelligan, 2013) and the head circumference is high (above the 90th percentile) (Wilbrand, Bierther, Nord et al., 2014).

2. Literature review

Environmental factors are identified as having a greater influence on the incidence of scaphocephaly than its genetic counterpart, which comprises about 2% of cases (Lajeunie, Merrer, Bonaiti-Pellie et al., 1996). Multiple gestations, large infant size, uterine abnormalities or abnormal intrauterine position are identified as risk factors in antenatal head compression, which is associated with non-syndromic craniosynostosis (Graham & Smith, 1980; Hunenko, Karmacharya, Ong et al., 2001). Higginbottom et al. further notes that external forces to the breech position of the head, the amniotic band and a morphologic abnormality of the uterus are also risk factors (Graham & Smith, 1980). Instances of twin studies show that the gestational constraints promote the premature sutural fusion in all twin types (Boyadjiev, 2007).

Other non-genetic risk factors include maternal smoking, advanced maternal age, white maternal race, use of nitrosatable drugs (such as nitrofurantoin), gestation at high altitude, paternal occupation (especially agriculture or forestry), endocrine abnormalities (e.g., hyperthyroidism), fertility treatments, as well as warfarin ingestion during gestation (Alderman, Lammer, Joshua et al., 1988; Kallen, 1999; Alderman, Zamudio, Baron et al., 1995; Gardner, Guyard-Boileau, Alderman et al., 1998; Bradley, Alderman, Williams et al., 1995).

Delshaw et al. delineated four concepts that are descriptive of the ensuing changes in the cranial vault expansion due to the diminished growth at the fused suture (Delshaw, Persing, Broaddus et al., 1989; Williams, Ellenbogen & Gruss, 1999). Namely, the premature fusion of the cranial

vault bones act as a single plate with decreased growth potential; the abnormal asymmetrical bone deposition takes place at the perimeter sutures with increased bone deposition directed away from the bone plate; the perimeter sutures adjacent to premature fused suture compensate in growth more than perimeter sutures distant to the sutural stenosis; and a non-perimeter suture that is neighbored to the premature fused suture undergoes enhanced symmetrical bone deposition along both edges.

Recent findings indicate that even for mild cases in non-syndromic synostosis a significant number exhibit intracranial hypertension (defined as $>17\text{mmHg}$) on lumbar puncture (Inagaki, Kyutoku, Seno et al., 2007). The increase in intracranial pressure (ICP) occurs because the cranial vault (compartment) is too tight to hold its contents (the brain). Furthermore, there may be an inconsistency between the cerebrospinal fluid production and its egress. The pressure on the brain signifies neurologic sequelae, which include headaches, nausea, vomiting, visual problems, motor and behavioural/intellectual delay.

The three principle objectives of surgical treatment for scaphocephaly are to prevent increased ICP, to allow normal brain development and to achieve a cosmetically acceptable craniofacial morphology or structure. The surgical correction serves to counteract the craniofacial skeleton aesthetic and functional anomalies in order to restore a normal spatial relationship between the skull and the contained neural structures (Ciurea, Toader & Mihalache, 2011).

For children diagnosed with scaphocephaly, surgery is often performed in their first year of life. It is found that it is usually these children who show significant mental improvements, rather than those who receive surgery when they are older as it results in minimal impairment of brain development and optimal bone regeneration (Renier, Lajeunie & Arnaud et al., 2000). Older patients with scaphocephaly are often at risk for intracranial hypertension (ICH) and require more extensive operations to achieve an aesthetic head shape.

Surgeries performed among children include the strip craniectomy, H-procedure, “pi” procedure, vertex craniotomies, subtotal or total cranial vault remodeling as well as helmet-molding and endoscopy-assisted techniques (Antunes, Arnaud, Cruz et al., 2009). The craniofacial surgical unit at Tygerberg Hospital modified the technique of total vault remodeling to improve the stability of the reconstruction, to increase the biparietal distance (which is decreased with scaphocephaly), to reduce operation time and lastly to avoid secondary procedures for the removal of titanium plates which are considered as rigid.

The decision to operate depends on the infants' appearance and functional concerns. Cases of mild deformity without clinical evidence of intracranial hypertension or developmental delays can be managed by observation. Those at risk with significant dysmorphology especially with the suggestion of increased brain pressure should be treated operatively (Steinbacher & Bartlett, 2013). The treatment goals are: (1) release the fused sutures to allow for brain growth and development; (2) normalize the head and forehead shape; and (3) mitigate functional issues (e.g., intracranial hypertension and developmental delay).

In the neonatal period, when the anteroposterior deformity may be mild, a simple “strip” craniectomy (sagittal synostectomy) or a variation thereof may be performed. This can be done by either a full open or an endoscopic approach. Cuts are made along the fused sagittal suture and sometimes additional cuts are made laterally into both parietal bones. The space created between parietal bones at the vertex may allow the head shape to normalize with brain growth (the fronto-occipital distance shortens and the biparietal distance expands) (Williams, Ellenbogen & Gruss, 1999). However, the frontal bossing or occipital prominence can persist over time. Additionally a fusion of the suturectomy borders could occur (recurrence of the scaphocephaly). This is not an attractive surgical option for infants older than 6 months because: (1) it does not adequately correct the scaphocephaly; and (2) permanent osseous defects can result along the vertex. It necessitates a period of a therapeutic or molding helmet use following surgery for an estimated 12-18 months. Therefore, this procedure can only be performed when the patient is younger, typically three months of age. Craniofacial surgeons have reported on

using endoscopic access for treatment of sagittal synostosis with various modifications (Teichgraeber, Baumgartner, Waller et al., 2009). It has been demonstrated that, in infants younger than 3–6 months of age, treatment of sagittal synostosis by suturectomy and parasagittal osteotomies can be effective.

The second type of surgery is a subtotal cranial reconstruction to contour the posterior two thirds of the skull. The forehead is typically left to remodel on its own. If the dysmorphology is more significant, but still minimal frontal prominence is present, the “pi” procedure can be performed. The “pi” procedure immediately corrects the fronto-occipital length and biparietal width (Jane, Edgerton, Futrell et al., 1978). Technically, it involves two parallel parasagittal osteotomies of the two parietal bones, connected with a transverse osteotomy, located behind the coronal suture and extends to the temporal region (resembling the Greek symbol for pi). The dura is dissected free from the endocranial surface of the frontal bone and the remaining fused sagittal suture, to prevent buckling when reapproximating new bone edges. Displacing the frontal bone posteriorly results in bulging of the brain laterally. These two movements are the key to improve the morphology of scaphocephaly. Though probably not more effective than strip craniectomy during the neonatal period, the “pi” procedure offers the advantage of immediate aesthetic correction of head shape and improved efficacy in older infants (e.g., 8 months).

Moderate to severe scaphocephaly is best corrected by total cranial vault reconstruction; as the more limited techniques described will not adequately correct the dysmorphology (Francel, 1995; Panchal, Marsh, Park et al., 1999; Weinzweig, Baker, Whitaker et al., 2002). This is best done between 6 and 9 months and entails excision of the frontal, parietal, and occipital bone plates, which are molded, reshaped, and repositioned. The occipital region is advanced forward and the frontal prominence retro-positioned. When performed in children older than 2 years, few if any calvarial defects are left, because they are less apt to reossify (Weinzweig, Baker, Whitaker et al., 2002).

The stabilization of repositioned bone segments is best performed with sutures, wires or plates and screws. The objective is to create a stable construct, but not to restrict brain growth and subsequent skull expansion. For the infants undergoing cranial remodeling, titanium or metallic plates and screws could be used. These plates need to be removed due to the possibility of transcranial migration with growth and they might possibly restrict the cranial growth. Free-floating forehead techniques have been attempted in the past, but without stability, a permanent deformity may result. Obviously, limited or endoscopic techniques do not utilize fixation, as they involve strip suturectomies and/or barrel staves only.

Looking at new ways to address the issues of surgical treatment, the three principle objectives of surgical treatment for scaphocephaly will need to be aligned to prevent increased ICP, allow normal brain development and to achieve an acceptable cranial morphology or structure.

3. Aim of the Investigation

The aim of this study is to evaluate the surgical outcomes of the modified total cranial vault remodeling procedure for the management of sagittal synostosis, which were performed over three-years from October 2011 to May 2014.

3.1 Objectives

The objectives of this study are:

- a. To evaluate the effectiveness of the onlay lag fixation technique with self-drilling screws as a method for internal fixation.
- b. To evaluate the effect of the modification on the surgery length and compare it to the traditional technique.
- c. To compare the average volume of blood transfusion for the traditional and modified technique.
- d. To clinically evaluate the effect of the modified technique on the progress of head growth by head circumference measurements.
- e. To evaluate parental satisfaction with the aesthetic outcome.

4. Methodology and Materials

A patient record review was undertaken, whereby the medical records database of the Division of Plastic and Reconstructive Surgery at Tygerberg Academic Hospital was searched to identify pediatric patients who underwent surgery with our modified technique of total cranial vault remodeling for scaphocephaly from October 2011 to May 2014. Patients' records review was the primary mechanism for collecting data. A standardized intake sheet (Appendix A) was administered for each case in order to capture the biographical details of the participants, along with the pathological features.

4.1 Selection and Description of Participants

The study sample comprised pediatric patients who underwent surgery for sagittal synostosis at Tygerberg Academic Hospital. Medical records of all pediatric patients who underwent sagittal synostosis were reviewed in their entirety.

The selection criteria are as follows:

4.1.1 Inclusion criteria

- a. Male and female children with isolated, single suture sagittal synostosis confirmed by computed tomographic scans.
- b. Pediatric patients who underwent reconstructive surgery, namely the modified total cranial vault remodeling procedure for the treatment of single suture sagittal synostosis.

4.1.2 Exclusion criteria

- a. Presence of major medical or neurological conditions (e.g. cardiac defects, seizure disorders, cerebral palsy, significant health conditions requiring surgical correction).
- b. Multiple suture craniosynostosis or the presence of two or more cranial malformations.
- c. Patients who underwent total cranial vault remodeling which was intended to address abnormal head shape resulting from the other fused sutures.
- d. Patients diagnosed with a genetic syndrome or syndromic sagittal craniosynostosis.

4.2 Traditional Surgical Technique

Traditionally the division of plastic and reconstructive surgery at Tygerberg Academic Hospital employed the total cranial vault remodeling technique as an ideal reshaping procedure to reassemble the cranial bone pieces to normalize the shape of the deformed skull.

The craniotomy lines and the bone pieces are marked as: (1) A for forehead; (2) the temporal and parietal bones are marked into pairs of bone strips as B and B', C and C', D and D', E and E' pieces for right and left side respectively; and (3) F and F' for the occipital bone piece. The forehead bone is displaced backward and fixed into the new position by wires. Then the rest of the reassembling follows by transpositioning the matched pairs of temporal and parietal bone strips from right to left and vice versa with a rotation at 90 degree (the sagittal border of the bone piece become temporal or parietal border and vice versa) to form the final remodeled shape of the head. The osteotomy bone plates are fixated to lateral skull bone edges by wires. For approximation and fixation of the bone pieces to each other, PDS sutures are used. The bone dust slurry is used to fill the gaps between bone pieces.

This assembling technique of transpositioning and shifting bone pieces results in transforming the head shape from high and narrow to a flatter and broader head shape. This promotes a normal head shape.

4.3 Modified Total Cranial Vault Reconstruction

In recent years, the technique was modified in order to improve the stability of the reconstruction to increase the biparietal distance (which is decreased with scaphocephaly), to reduce operation time and, lastly to avoid secondary surgical procedures whereby the rigid titanium plates need to be removed.

The modified technique is performed through a transcoronal zigzag incision and exposure of the cranial vault by elevating the scalp at the subperiosteal level as one flap that includes the pericranial layer. After the exposure and haemostasis, the craniotomy design will be marked. According to the size of the child's head, an ideal bone plate is chosen to construct the planned new forehead by using a forehead template. Then the new forehead replaces the old forehead, is

repositioned backwards in a posterior direction to improve frontal bossing, and is fixed by using two-hole resorbable plates with self-drilling titanium screws (DePuy Synthes).

The rest of the total remodeling is done by reassembling the rest of the removed bone plates combined with a barrel stave procedure at the occipital side of the skull to recreate a normal head shape. Instead of fixating the craniotomy pieces to the skull bone by laying them edge to edge and fixing them by using plates and screws in the temporal and parietal area, we implemented the onlay lag fixation technique by overlying the craniotomy bone pieces on the temporal and parietal skull bones and fixating with 4-8 mm long self-drilling screws. A lag technique is applied to obtain the compression effect of the screws on the overlying bone pieces creating enough stability at the fixation point. The outer bone piece hole is over-drilled by the size of the external diameter of the screw thread to form a gliding hole to mimic the effect of the true lag technique. This fixation is of advantage for cranial growth, as the screws are fixated at one point only and not two points of fixation as with plates and wires, which could lead to growth restriction. Furthermore, the bone plates can function like a small hinge. The biparietal distance is increased through the onlay technique depending on the bone thickness between 3 to 10mm over a broad surface area. The bone dust slurry and small pieces of bone that are left are used to fill the gaps. We use fibrin glue to stick the bone dust slurry and small bone pieces to the exposed dura.

Closure of the wound over one small suction drain is done by subcutaneous 3/0 Vicryl and for the skin by 4/0 and/or 3/0 Vicryl rapid sutures. Dressing is done by a ribbon band soaked with Friar's Balsam (Tincture of benzoin solution), which is fixed with staples over the wound.

Resorbable plates and titanium self-drilling screws for fixation of the reconstructed forehead are used to prevent growth restriction. According to the supplier, the resorbable plates are expected to be absorbed within a twelve month period.

4.4 Data Collection Method

Patients attended follow-up visits and their progress was observed. The medical records for patients undergoing the above-mentioned technique were retrieved and served as secondary data for the purpose of analysis. Measurements were taken by means of the patients head circumference (HC).

Medical records with surgical reports, history, as well as pathological features were reviewed using a standardized intake sheet (Appendix A). The selected records are presented along with a descriptions of their clinical manifestations/presentation, neuroimaging and mid-term outcomes.

Factors studied included: a) patient demographic characteristics; b) duration of surgical procedure; c) pre-, intra- and post-operative haemoglobin levels and blood transfusion amount; d) length of Intensive Care Unit (ICU) and hospital stay; d) presence of intracranial hypertension (ICH); e) postoperative complications; f) parents satisfaction and g) changes in HC measurements.

4.4.1 HC Measurement Procedure

The HC of the children was measured using a flexible non-stretchable measuring tape. The HC measuring tape was placed around the child's head so that the tape lied across the frontal bones of the skull, slightly above the eyebrows, perpendicular to the long axis of the face, above the ears and over the occipital prominence at the back of the head. The tape was moved up and down over the back of the head to locate the maximal circumference (National Health & Nutrition Examination Survey, 2012).

The process was repeated twice for each subject before and after surgery. The measurements for the HC were plotted and interpreted for the children using the Standard Infant HC for age growth charts (Appendix B and C). The HC for age percentile interpretation was used to define the HC: Percentile < 3 is regarded as microcephaly; percentile > 3 and < 97 is seen as normal HC; Percentile > 97 is regarded as macrocephaly. For children who were older than five years of age

a different head circumference –for age growth chart was used (Tanner & Whitehouse, 1973; Appendix D).

The aesthetic outcome of surgery was also evaluated. The outcome was measured by using a scale from 1 to 10 to express the parents' satisfaction level (from 1-3 is poor, 4-7 is satisfied and 8-10 is good).

4.5 Ethical considerations

This research falls under the ‘Human subject research’ since data was collected pertaining to an individual’s medical condition, treatment thereof and outcome. The four ethical considerations that were undertaken in this study are the protection of participants from harm (physical and psychological), prevention of deception and confidentiality. The responsibility of the investigator to the participants is to ensure that anonymity, avoidance of harm, reciprocity and dissemination of the results. The collected secondary data was kept in a secure location to which only the investigator (conducting the research study) has access to.

This retrospective study received ethical approval (ethics reference number: S14/07/144) and was conducted in accordance to ethical guidelines as prescribed by the Ethical Committee of University of Stellenbosch, South Africa.

4.5.1 Confidentiality and Management of Data

Information about study subjects will be kept confidential and managed according to the requirements of the Health Research Ethics Committee at the University of Stellenbosch. Patient data will be entered into electronic spreadsheets. One spreadsheet (the correlation tool) will contain the patient name, medical record number, and patient study number. The second spreadsheet will contain the patient study number, as well as all of the variables required by the study. The two spreadsheets will be stored as separate files, protected by unique passwords. Only the investigators will have access to the files and their passwords. Any paper records will be stored in hard copy in a locked filing cabinet in the investigator’s office for a minimum of five years.

5. Results

Ten patients were identified from the database that had sagittal synostosis and had undergone a cranial vault remodeling procedure at an age greater than 6 months. The medical records of two patients were unavailable and thereby excluded. The remaining eight patients' ages ranged between 4 months to 5 years and 7 months with an average of 1 year and 11 months at presentation.

The patients comprised of three boys and five girls. All the patients underwent the modified single-stage total cranial vault procedure. The patients' average age at the time of cranial vault remodeling was 2 year and 10 months (range 8 months to 5 years and 9 months). Only one patient presented with a raised intracranial pressure, which was treated by shunting and needed to be revised during the scaphocephaly surgical repair.

The preoperative haemoglobin levels were comparable at an average of 10.1g/dL. All the patients' intraoperative haemoglobin levels were above 7 g/dL, with an average of 8.4g/dL. All patients showed a higher haemoglobin level post-surgery with an average of 11.3g/dL. Intraoperative blood transfusion (packed red blood cells) was used for 7 patients with an average volume of 230 ml. One of the patients received postoperative transfusions.

Procedures were completed with an average surgical time of 3.4 hours and ranged between 2.58 – 4.5 hours. The average hospital length of stay for the eight patients was 8.38 days (ranging from 7 to 10 days), with an average ICU stay of 1 day across all participants.

The parents' satisfaction yielded an average of nine out of 10 which expressed their satisfaction with the aesthetic outcome of their children's improved head shape. The parents of a child with a visible hairless scar rated the outcome as 6 out of 10 on the satisfaction score.

The average follow-up period was 1.01 year (ranging from 7 months to 1 year and 9 months). All the operations were performed by the division of plastic and reconstructive surgery in cooperation with the division of neurosurgery at Tygerberg academic hospital.

5.1 Head Shape

Pre- and post-operative head circumference scores were plotted for seven patients using the head circumference-for-age growth chart (WHO) for boys and girls under 5 years of age (Appendix B and C). One patient was scored as per the Tanner and Whitehouse (1973) head circumference growth chart as his age was beyond five years. In all patients, the post-operative head circumference percentile measurement was higher than pre-operative.

Results showed that three of the five females have post-surgical HC readings above the 97th percentile; one female scored on the 85th percentile the other one between the 50th and 85th percentile. For the three boys, two scored post-surgical HC readings above the 97th percentile and another above the 10th percentile.

87.5% of the patients' results show that post-surgical head circumference measures are above the 50th percentile on the growth charts (63% of patients were above the 97th percentile, 12.5% of patients were above the 50th percentile and 12.5% was above 85% percentile and 12.5% above the 10th percentile).

5.2 Complications

One patient had a sterile fluid collection around the plates, which required drainage. After the one-year follow-up period, one patient developed total fusion of all cranial sutures of the remodeled cranial vault bones (pansynostosis) with raised intracranial pressure, which was independent of the scaphocephaly repair procedure. Reoperation was necessary, which showed that the screws were covered by new bone formation. At the time of follow-up, three patients showed that the resorbable plates failed to be resorbed timeously and required removal.

One patient was readmitted to the hospital because of wound infection and dehiscence, which was treated conservatively by dressing and oral antibiotics without operative intervention. Scar alopecia was present in one patient.

6. Discussion

Surgical correction of scaphocephaly should lead to significant improvement in the head shape over the months following surgery. The parents should be given a realistic appraisal of the benefits of surgery from both an appearance and functional point of view. They should be counseled that a tendency toward recurrence of the synostotic morphology might occur secondary to diminished growth in the region. Therefore, the child's deformity is often operatively overcorrected (Steinbacher & Bartlett, 2013). Additionally, secondary deformities (e.g., temporal hollowing), also from a lack of growth, may pose an aesthetic concern to the child for several years postoperatively, requiring secondary extracranial intervention. Repeat intracranial procedures may rarely be required (Steinbacher & Bartlett, 2013).

The options for treating sagittal suture fusion include simple “stripcraniectomy” (2cm), extensive craniectomies (6-8cm) and craniotomies combined with reconstructive procedures (cranial vault remodeling) (Ciurea, Toader & Mihalache, 2011). In general, the technical surgical goals for the cranial vault remodeling entail: releasing the area of sutural fusion, repositioning the bone in an anatomic location, eliminating secondary compensatory changes, filling in osteotomy gaps with bone dust slurry, and closing the soft tissue relatively tension-free.

The division of plastic and reconstructive surgery at Tygerberg academic hospital adapted the total cranial vault remodeling procedure as the first choice of scaphocephaly repair with forehead reconstruction. The unit's old technique aimed to improve the scaphocephalic head shape by transposition and shifting bone strips of the cranial vault bones. This technique relied on the use of wires and sutures for the fixation of the bone pieces to each other and to the skull. Recently, we adapted the following modifications of the old technique: 1) We mark and design a new forehead using the forehead template on the cranial vault bone. This ideal new forehead is then repositioned at a posterior location and fixated. 2) For providing more intracranial volume, we developed a technique of fixing the strips of the cranial vault bones to the skull by overlapping the bone edges (onlay lag fixation technique). Depending on the bone thickness (3 to 10 mm),

this increases the biparietal distance over wider areas. It also functions as a hinge fixation point to adapt the increasing size of the brain after the craniotomy.

Another disadvantage of the previous technique was the possibility of sinus formations caused by the wire fixation. This necessitated frequent interventions.

With the old technique, the length of the surgical procedure (ranging from five to six hours) exposed the child to the risk of the side effects of the long general anesthesia and subsequent longer ICU stay of two to three days post-surgery.

However, with the modified technique, the surgical time was reduced to an average of 3.4 hours that resulted in the minimization of the overall risk of the side effects of the long anesthesia and shortening of the overall hospital stay including the ICU stay - the average ICU stay was one day and the hospital stay 8.38 days. Regarding the blood transfusion, an average of 230ml was used with the modified technique in comparison to the 763ml of the old technique.

Other studies show an average surgical time of 5.4 hours. Furthermore, they show an average hospital stay of 4.1 days, an average ICU stay of 1.3 days, and an average transfusion volume of 423ml (Rottgers, Kim, Kumar et al., 2011).

Unexpectedly, the resorbable plates were clinically observed and palpable after 12-months. Also, one of the patients developed a fluid collection with bone absorption and a visible cosmetic defect, which required a secondary surgical procedure to remove the plates. This led to the omission of the plates for future use. Subsequently, titanium plates and screws are used and removed routinely after three months.

Contrary to the literature, seven of the eight patients had a head circumference pre-operative below the 97th percentile. The head circumference was below the 50th percentile in six of the eight patients. No explanation for this deviation from previously described scaphocephalic head shape characteristics can be given.

The complications associated with the modified procedure are substantial, but comparable to the traditional method.

Limitations of the Study. The study sample size is too small to make further inferences. The loss of medical reports due to an improper hospital archiving system is of limitation as well. In addition, a standardization of measurements is inaccessible due to the unavailability of pre-operative cephalic index measures in the patients' medical reports.

Implications for further research or practice. There is a need for larger sample of patients along with longitudinal investigations to monitor patient outcomes following surgery. The implementation of the cephalic index measurements for pre- and post-operative evaluations to standardize the patients' outcome data is important to make the data comparable to other investigations. Furthermore, there is a need to raise awareness about the early diagnosis of craniosynostosis and its appropriate referral pathways by educating health professionals at primary health care facilities.

7. Conclusion

The onlay lag fixation technique using self-drilling screws provides adequate stability for the new remodeled bone plate. It also increases the biparietal distance by the thickness of the bone in the temperoparietal area as well as the head circumference. In addition, the one point fixation concept works as a hinge mechanism in the biparietal direction should the brain need more space.

Relative to the traditional technique for scaphocephaly repair, the modified technique can be safely applied for a wide range of children and yields good parents' satisfaction. Furthermore, it reduces the need for blood transfusion by two-thirds and can be performed faster, which has implications on the patient's recovery period and the cost of the procedure.

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Appendix A**Standard Intake Sheet: Patient Data**

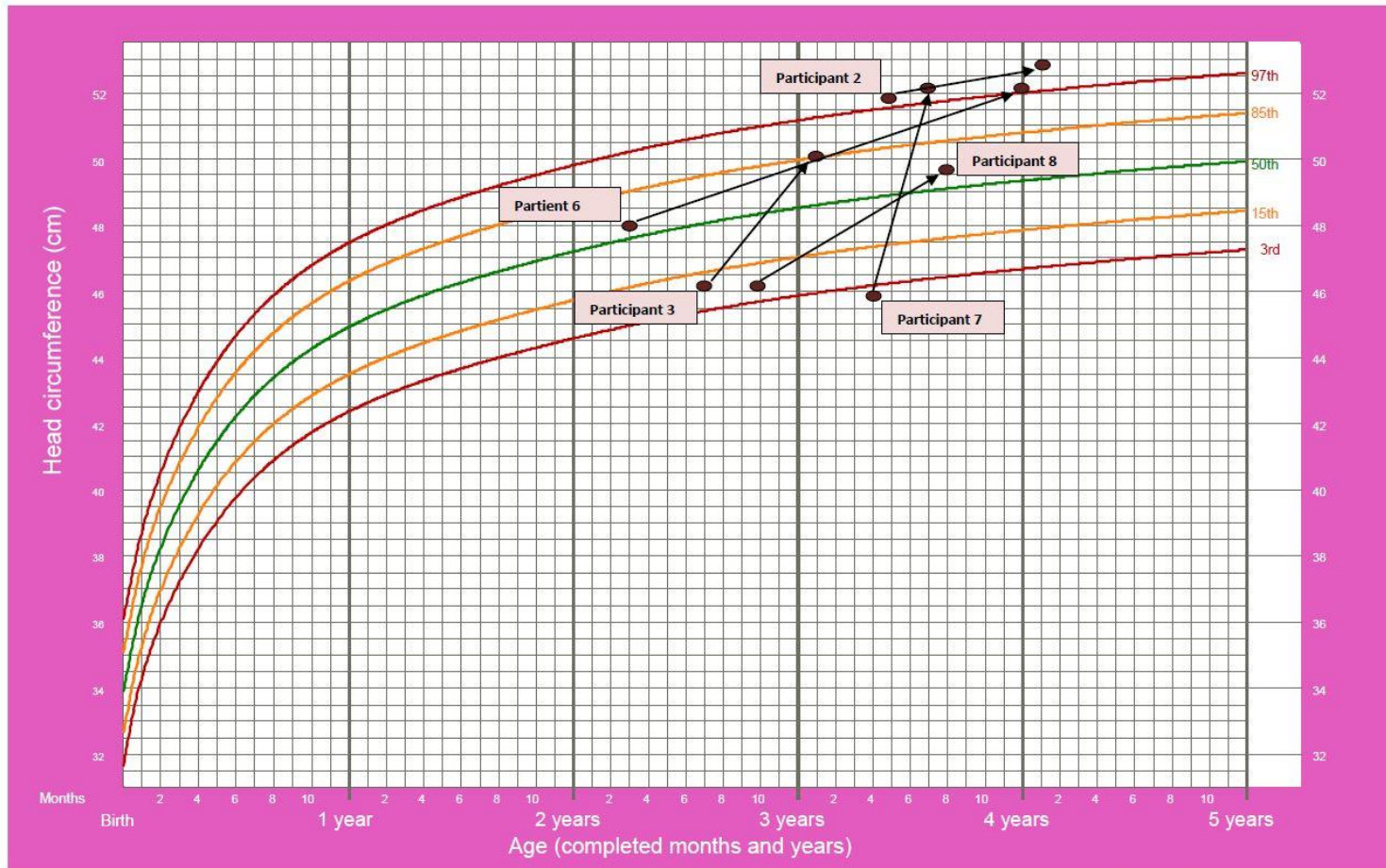
1. Name of patient		
2. Sex	Male:	Female:
3. Method of Birth:	C-Section Surgery:	Vaginal Delivery:
4. Age at Birth	(weeks)	
5. Age at Birth	(grams)	
6. Head Circumference at Birth	(cm)	
7. Age at Presentation	(years)	
8. Age at Surgery	(months/years)	
9. Head Circumference before Surgery	(cm)	
10. Pre-Surgery Haemoglobin Level	(gm/dl)	
11. Duration of Surgical Procedure	(min)	
12. Intraoperative Haemoglobin Level	(gm/dl)	
13. Blood Transfusion	(ml)	
14. Intensive Care Unit (ICU) Stay	(day/s)	
15. Hospital Stay	(day/s)	
16. Haemoglobin Levels on Hospital Discharge	(gm/dl)	
17. Intracranial Hypertension	Yes:	No:
18. Head Circumference	(cm)	

following Surgery										
19. Mother's Satisfaction	1	2	3	4	5	6	7	8	9	10
20. Follow-up since Surgery	(years)									
21. Post-surgical complications										

Appendix B

Head circumference-for-age GIRLS

Birth to 5 years (percentiles)

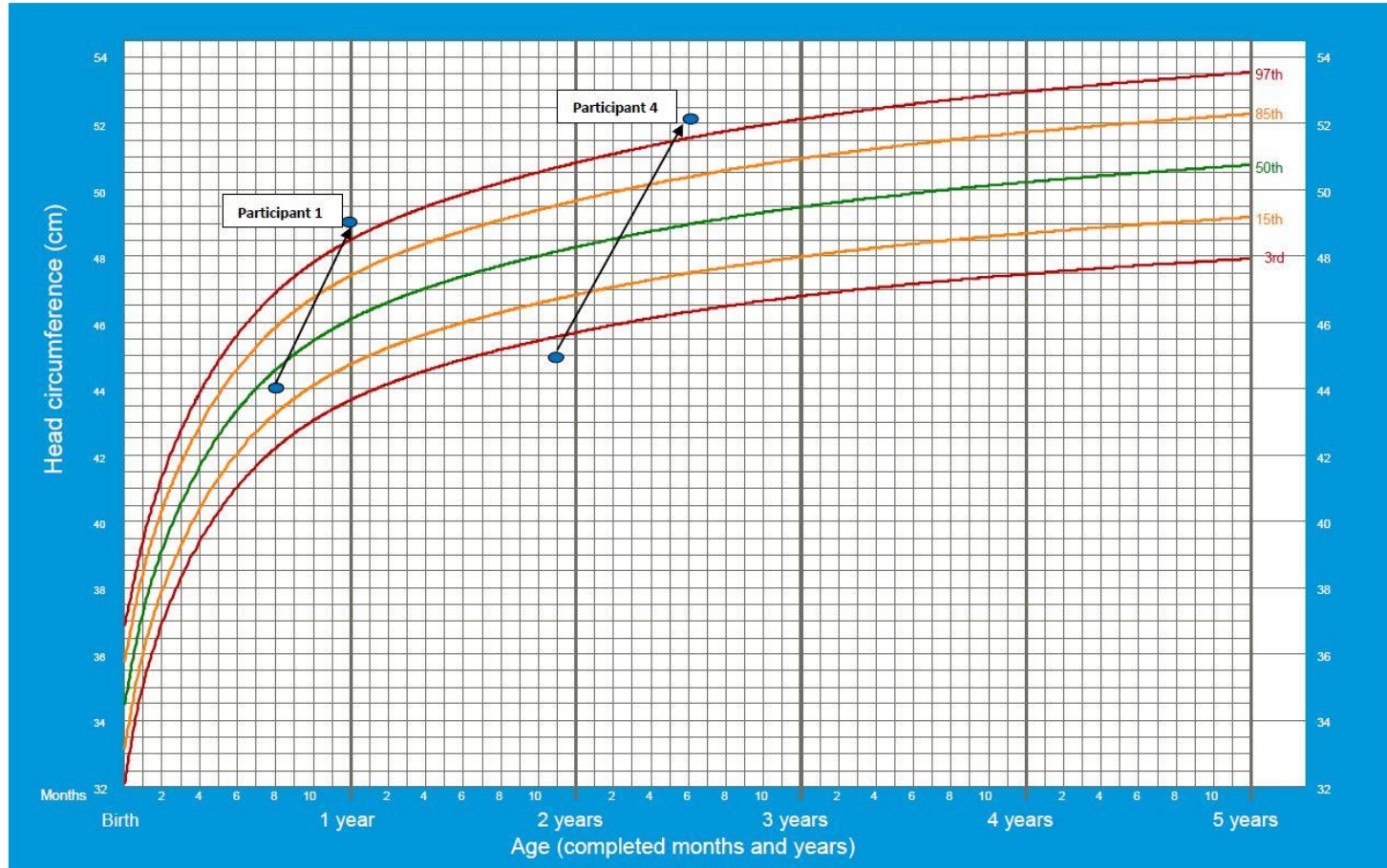


WHO Child Growth Standards

Appendix C

Head circumference-for-age BOYS

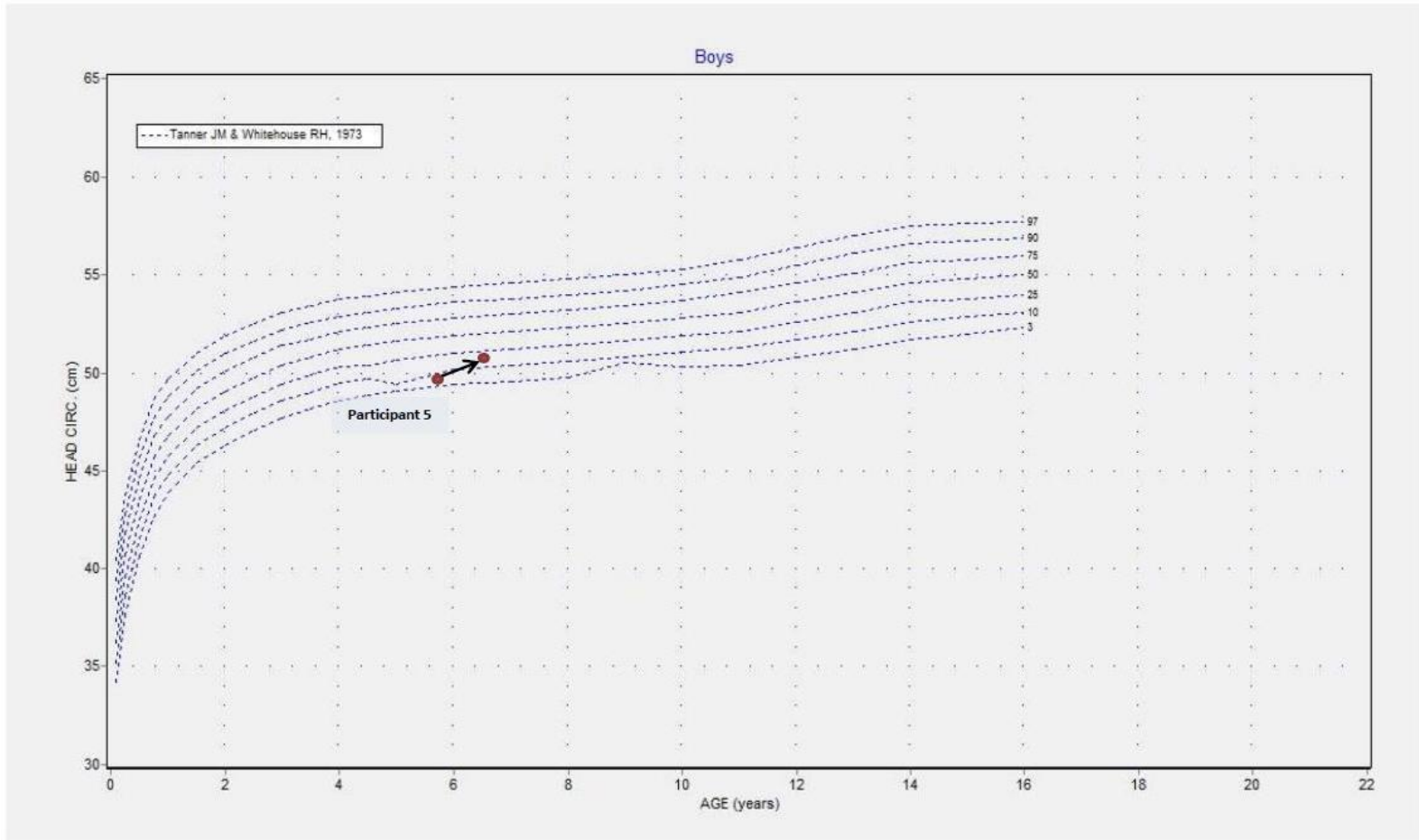
Birth to 5 years (percentiles)



WHO Child Growth Standards

Appendix D

Tanner and Whitehouse Head Circumference for Age Growth Chart (boys from 0-18 years)



Appendix E

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7	Patient 8	Patient 9	Patient 10
Sex	Male	Female	Female	Male	Male	Female	Female	Female		
Method of birth	Vaginal Delivery	C- Section	Vaginal Delivery	C- Section	Vaginal Delivery	Vaginal Delivery	C- Section	C- Section	C- Section	Vaginal
Weight at birth (grams)	3110	1200	920	3410	1152	1340	1230	655	325	
Age of birth (weeks)	38	32	26	40	37	32	33	27	40	
Age of Presentation (years)	0.33	3.17	1.83	0.67	5.58	1.42	1.42	1.17		
Age of Presentation (months)	4	3 yrs 2	1yr 10	8	5yr 7	1yr 5	1yr 5	1yr 2		
Age at surgery (years)	0.67	3yr 5	2.58	1.92	5.75	2.25	3.33	2.83	13	
Age at surgery (months)	8	41	2yr 7	1yr 11	5yr 9	2yr 3	3yr 4	2yr 10		
Hae mogl Pre-surgery (g/dL)	9	14	8	11	11	11	9	8		

	Intraoperative (g/dL)	8	8	9	9	8	8	8	9		
	Post-operative (g/dL)	13	12	12	11	12	10	9	11		
Duration of surgical procedure (hours)		3.78	2.58	2.83	3.08	3.75	4.5	3.5	3.5		
Blood transfusion (ml)		390	150	300	300	200	250	250	0		
ICU stay (day)		1	1	1	1	1	1	1	1	1	
Hospital stay (day)		10	9	8	8	7	9	8	8	8	
Intracranial Hypertension		No	No	Yes	No	No	No	No	No		
Head Circumference	At birth (cm)	33	43	46	35	27	40	40	24	36	
	Before Surgery (cm)	44	52	46	45	49	48	46	46		
	Following Surgery (cm)	49	53	50	52	51	52	52	49	52	

Follow-up period since surgery (years)	1	0.67	1.33	0.58	0.75	1.75	1.25	0.83		
Follow-up period since surgery (years)	1yr	8	1yr 4	7	9	1yr 9	1yr 3	10		
Age at follow-up (years)	1yr 8	4yr 1	3yr 1	2yr 6	6yrs 6	4yrs	3yr 7	3yr 8		
Parents Satisfaction	10	10	8	10	9	10	10	6	9	