

Original Article

Endoscopy-assisted craniosynostosis surgery followed by helmet therapy

H. H. K. Delye, W. A. Borstlap¹, E. J. van LindertDepartment of Neurosurgery, ¹Department Oral and Maxillofacial Surgery, Radboudumc Nijmegen, The NetherlandsE-mail: *H. H. K. Delye - Hans.Delye@radboudumc.nl; W. A. Borstlap - Wilfred.Borstlap@radboudumc.nl; E. J. van Lindert - Erik.vanLindert@radboudumc.nl

*Corresponding author

Received: 15 January 18 Accepted: 18 January 18 Published: 07 March 18

Abstract

Background: Surgical methods to treat craniosynostosis have evolved from a simple strip craniectomy to a diverse spectrum of partial or complete cranial vault remodeling with excellent results but often with high comorbidity. Therefore, minimal invasive craniosynostosis surgery has been explored in the last few decades. The main goal of minimal invasive craniosynostosis surgery is to reduce the morbidity and invasiveness of classical surgical procedures, with equal long-term results, both functional as well as cosmetic.

Methods: To reach these goals, we adopted endoscopy-assisted craniosynostosis surgery (EACS) supplemented with helmet molding therapy in 2005.

Results: We present in detail our surgical technique used for scaphocephaly, trigonocephaly, plagiocephaly, complex multisutural, and syndromic cases of craniosynostosis.

Conclusions: We conclude that EACS with helmet therapy is a safe and suitable treatment option for any type of craniosynostosis, if performed at an early age, preferably around 3 months of age.

Key Words: Craniosynostosis, endoscopy, helmet, minimal invasive, surgical technique

Videos available on: www.surgicalneurologyint.com
Access this article online
Website: www.surgicalneurologyint.com
DOI: 10.4103/sni.sni_17_18
Quick Response Code:


INTRODUCTION

The history of the identification of different types of craniosynostosis, the underlying pathogenesis, and the subsequent development of surgical treatments for this entity reads as a very entertaining novel. In the last decade, many reports have reviewed the history, treating paradigms, and evolving surgical techniques in much detail.^[3,26,30,32,34] In general, surgical techniques have always reflected the contemporary beliefs about pathogenetic paradigms and have always been limited by the available (or missing) technology, regarding both surgical tools and anesthesia.

At the time Virchow stated his law, it was believed that the observed deformities in craniosynostotic skulls were

a result of cessation of growth across a prematurely fused suture, with compensatory growth along nonfused sutures in a direction parallel to the affected suture, causing obstruction of normal brain growth.^[40] Hence, the first surgical attempts to treat this condition in the late 1800s

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Delye HH, Borstlap WA, van Lindert EJ. Endoscopy-assisted craniosynostosis surgery followed by helmet therapy. *Surg Neurol Int* 2018;9:59. <http://surgicalneurologyint.com/Endoscopy-assisted-craniosynostosis-surgery-followed-by-helmet-therapy/>

consisted of suturectomy,^[23,24] although it appears that many children treated at that time were more likely to have microcephaly rather than craniosynostosis. This distinction was not diagnosed or considered at that time. As patients were only treated when neurological deficits developed, these procedures were performed at an older age and frequently reossification occurred before correction of the skull shape was established. The outcome of these early procedures was not satisfying and a high mortality rate was associated with these procedures, leading to fierce resistance by Jacobi and discontinuation of this technique.^[13] Some decades later, Faber and Towne reported excellent preservation of neurological function with minimal morbidity and mortality by performing suturectomy for craniosynostosis, presumably well differentiated from microcephaly.^[8] By the 1940s, strip craniectomies were widely accepted and it became clear that early intervention – at that time described as the period before 2 months of age – led to better functional as well as cosmetic outcome, a parameter that was not of primary importance at that time.^[9] However, the problem of reossification in older children remained and required extensive and difficult secondary cranial reconstruction operations, associated with high morbidity and mortality rates. Therefore, several techniques were developed to fight this reossification process, including wrapping of the cut bone edges with polyethylene or tantalum foil or applying Zenker's solution to the dura.^[1,12,37] Wrapping the bone edges led to infections and still early reossification, Zenker's solution caused seizures, hence these techniques were discarded.

By the mid-1950s, there was a significant advance in anesthesia and blood transfusion and surgery for craniosynostosis became very safe. At that time, Moss rejected the Virchow's law and proposed his "functional matrix theory," stating that the active growth of the underlying brain dictated the passive cranial growth along the suture lines. He proposed that the cranial base and not the suture was the primary site of abnormality, with suture fusion being a secondary consequence.^[28] These beliefs, together with the technological advancement and clear failure of simple strip craniectomy procedures in older patients, led to the development of more extended procedures, in which for the first time cosmesis was considered as a primary indication for surgery by Shillito and Matson.^[36] From the early 1960s to mid-1990s several extensive calvarial remodeling techniques were developed.^[10,14,38] Tessier introduced pioneering techniques for the treatment of craniosynostosis that led to significant improvements in cosmetic outcomes, particularly for those with facial abnormalities.^[38] The limitations of suturectomy for advanced disease and the discovery by Delashaw *et al.* that a major cause of the cranial deformity was compensatory overgrowth at adjacent sutures, led to techniques in which the desired

changes in the shape and volume were established intraoperatively and the bony segments were fixed to maintain the correction.^[5] The outcomes of these techniques do not depend on postoperative brain expansion and are therefore more predictable than simple or extended craniectomy procedures. Therefore, cranial remodeling became the preferred surgical technique for craniosynostosis, although these techniques were associated with significant operative time, hospital stay, ICU monitoring, blood loss requiring transfusion, and complications.^[36] These limitations, especially blood loss and transfusion, were the motor for the development of minimal invasive craniosynostosis surgery techniques, using endoscopes or springs.^[19,25] In the early 1990s, Jimenez and Barone presented their minimal invasive suturectomy via endoscopic approach, supplemented with orthotic helmet molding therapy to treat scaphocephaly.^[19] As their experience grew, subsequent reports noted significant reduction in blood loss and need for transfusions, shorter operative times and hospital stays, decreased hospital costs with good to excellent cosmetic results, not only for scaphocephaly but also for trigonocephaly, anterior plagiocephaly, brachycephaly, and multisutural craniosynostosis.^[16-23] Several other groups have adapted these techniques and confirmed their findings.^[3,4,6,30,31,35]

The main goal of minimal invasive craniosynostosis surgery is to reduce the morbidity and invasiveness of classical surgical procedures, with equal long-term results, both functional and cosmetic.^[6,29,32]

Reducing the morbidity and invasiveness can be achieved by minimizing skin incisions and tissue dissection using the smallest working space possible while keeping good visual control over the surgical field to prevent major blood loss and other complications such as dural tears.

To reach these goals, we introduced endoscopy-assisted suturectomy (ECAS) supplemented with helmet molding therapy in our centre in 2005 and gained extensive experience with this technique.^[6]

METHODS

Surgical technique

General principles and equipment

This type of surgery can be performed with a standard armamentarium including the use of an endoscope with footplate and can be considered as a simple and easy surgery when performed correctly.

To minimize blood loss, we infiltrate the skin with lidocaine 2% or epinephrine 1:100.000. After skin incisions are made, we use monopolar cutting for galea and periosteum. The craniectomy is then initiated with a high-speed drill and continued with different rongeurs and Kerrisons. Any bleeding during surgery from the

epidural space and bone edges is easily controlled with FloSeal® Matrix Hemostatic Sealant and Osteone® bone wax (Baxter Healthcare Corporation, Fremont, CA, USA).

Once a small entrance craniectomy is performed, we use a 0-degree Storz lens scope with a working shaft used for endoscopic facial lift surgery without irrigation or suction to perform dura dissection from the overlying bone and synostotic suture [Figure 1]. This is usually very easy as the dura mater is hardly attached to a synostotic suture, but can be tricky in case of a deep and sharp bony ridge as is often the case in trigonocephaly. Blood aspiration is performed by a separate aspirator placed parallel to the endoscope. The endoscope with footplate allows good visual control of the operative field under the bone, identification and bipolar coagulation of transgressing emissary veins before rupture, and protection of the dura mater during bone resection. Craniectomy is then continued along the length of the affected suture under direct visual control of the endoscope. No subdural/subcutaneous drains are used and a small compressive head bandage is used for 24 h to prevent subcutaneous hematoma development.

Standard anesthetic monitoring techniques including electrocardiography, noninvasive blood pressure monitoring, pulse oximetry, temperature monitoring, and blood loss monitoring are used. As blood loss and operative times are very limited (30–60 min), there is no need for a central venous line nor an arterial line. Two peripheral venous lines suffice in all cases. Antibiotic prophylaxis consists of 25 mg/kg cefazolin i.v. given 20 min before skin incision. Postoperative monitoring is performed in pediatric medium care unit, with hemoglobin/hematocrit levels controlled 6 h after surgery and before dismissal the next day. Postoperative pain is treated with prophylactic paracetamol and low-dose i.v. morphine which can be tapered during the night after surgery. Because of the low level of morphinoids postoperatively and the very limited blood loss, there is no need for an urinary catheter during or after surgery. Patients can be orally fed 3–4 h after surgery. None of our patients need ICU monitoring postoperatively and



Figure 1: Instruments commonly used in EAC surgery. A: bone cutting scissors, B: small suction device, C: bended spatula for dura dissection, D: 0 degree endoscope with footplate

almost all patients are dismissed the day after the surgery. Helmet therapy is started 2 weeks postoperatively.

Jimenez and Barone showed that the critical age for EACS seems to be 6 months.^[22] After that age, the cosmetic results become worse and insufficient correction of skull shape is reached. However, one should not wait until the infant reaches the age of 5 or 6 months. The earlier an EACS is performed, the better the result. This is even more important in case of plagiocephaly and trigonocephaly. Therefore, below the age of 4 months, we always offer EACS as the treatment of choice, but for infants of 5 or 6 months, we restrict EACS for mild and moderate cases and consider open remodeling procedures for severe cases. We think that the optimal age to perform EACS is 3 months, as has been reported by other groups in the literature.^[3,30,31,35] At this age, the child has grown and acquired some weight after birth, and both preterm and term infants have recovered from the physiological anemia which is most severe at approximately 8 to 12 weeks after birth in term infants. In preterm infants who are already born with a lower hematocrit, this decline, referred to as anemia of prematurity (AOP), occurs earlier and is more pronounced in its severity than the anemia seen in term infants. Therefore, at the age of 3 months, the child can tolerate some moderate blood loss and is able to tolerate the molding helmet. Unfortunately, we are often confronted with a diagnostic/referral delay by general practitioners and pediatricians, because of which patients are only presented to us at a later phase, often after the age of 3 months.

In syndromic cases, we aim for very early surgery at an age of 4–8 weeks, as we try to halt the progressive deformity, prevent intracranial hypertension, and simplify reconstructive surgery at a later stage. Parents are informed that cranial vault expansion and bifronto-orbital advancement procedures will still be required at a later stage. Because of the very young age and additional problems such as sleep apnea and risk of increased ICP, molding helmet therapy has not been added in these cases up to now.

Scaphocephaly

Patients are positioned in prone sphinx position, aligning the sagittal suture with the horizontal plane [Video 1]. Two skin incisions of approximately 4 cm are used: one 2–3 cm behind the most posterior point of the anterior fontanel, and the second one 2–3 cm anterior of the posterior fontanel. From this skin incision, an osteoclastic craniectomy towards the anterior fontanelle and posterior fontanelle is performed using the high-speed drill and rongeurs after dissection and elevation of the periosteum [Figure 2]. The length of this craniectomy can vary in case a part of the suture is still open and patent. After this, FloSeal® Matrix Hemostatic Sealant is administered for hemostasis.

Then, the endoscope is introduced through the anterior skin incision and dura dissection from the overlying bone is performed. The perfect visualization of the dura and operative field by the endoscope in conjunct with a parallel positioned aspirator to clear any blood gives the surgeon total control of the operative field during this phase. Typically, in the middle to posterior part of the synostotic suture, several bridging veins running from the dura towards the bone can be identified and coagulated before rupturing. Hence, blood loss can be minimized and there is always perfect visualization of the dura and the underlying superior sagittal sinus. Once the dura dissection is completed, the periosteum is dissected and lifted from the suture. Bended bone cutting scissors are used to cut the bone strip from the posterior incision to the front, while the endoscope is used from the anterior incision to visualize and control the direction of cutting, protecting the underlying dura with the footplate. The removed bone strip should measure 4–5 cm wide. At this point, again FloSeal® Matrix Hemostatic Sealant is used covering the subcutaneous surgical field. Then wedge-shaped osteotomies are performed behind the coronal sutures and in front of the lambdoid sutures to assist in allowing an increase of the biparietal width. Periosteum, subcutis, and cutis are closed in separate layers using resorbable sutures, Steristrips™ (3M™, Diegem, Belgium) included. A small compressive head bandage is used for 24 h.

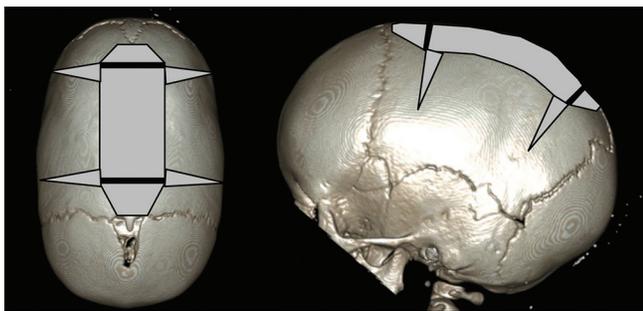


Figure 2: 3D scan showing extent of craniectomy in scaphocephaly. Thick black line indicates skin incision, grey area depicts craniectomy size

Trigonocephaly

Patients are placed in a supine position, aligning the metopic suture with the horizontal plane. One skin incision of approximately 3 cm is positioned symmetrically over the metopic suture just behind the hairline [Figure 3]. The exact position of this incision depends heavily on the preoperative 3D CT scan and is always a trade-off between the (future) hairline and the curvature of the forehead. When the hairline demands an incision that is not favorable to overcome the curvature of the forehead with the endoscope, we recently started to use a small zig-zag incision. This allows more anterior displacement of the skin, and thus, a more anterior entrance to the epidural space with the endoscope and less difficulties in reaching the endpoint of the craniectomy just above the nasion.

From this skin incision, an osteoclastic craniectomy towards the anterior fontanel is performed using the high-speed drill and rongeurs after dissection and elevation of the periosteum. The length of this craniectomy can vary in case a part of the suture is still open and patent. After this, FloSeal® Matrix Hemostatic Sealant is administered for hemostasis. Then, the endoscope is introduced and dura dissection from the overlying bone is performed. The perfect visualization of the dura and operative field by the endoscope in conjunct with a parallel positioned aspirator to clear any blood allows a safe dissection of the dura without the occurrence of dural tears although the frontal bone and synostotic suture often present with deep and sharp bony ridges. Typically, some bridging veins can be found near the most anterior part of the synostotic suture and can be coagulated before rupture. When performing dura dissection, the rigid scope tends to compress the dura as dissection advances anteriorly. Of course, it is of paramount importance to avoid too much pressure on the dura. Therefore, the entrance to the subdural space with the endoscope should be as anterior as possible to overcome the curvature of the forehead. Sometimes this will demand simultaneous progressive craniectomy of the suture while performing the dura dissection. Once the dura dissection is completed, the periosteum is dissected and lifted from the suture. A triangular craniectomy is

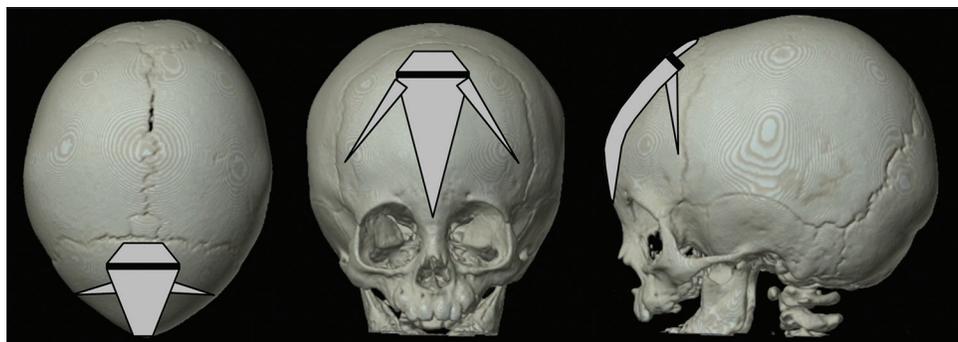


Figure 3: 3D scan showing extent of craniectomy in trigonocephaly. Thick black line indicates skin incision, grey area depicts craniectomy size

performed with a base of 3 cm, tapering down between the orbits to just above the nasion, using the endoscope to protect the dura and provide good visualization. The bone near the skull base is generally more thick and cancellous, causing more venous bleeding. This can easily be controlled by using FloSeal® Matrix Hemostatic Sealant, and Ostene® bone wax. We recently added small wedge-shaped osteotomies pointed towards the upper lateral orbital edges to our treatment protocol. We think this might assist in the outbending of the flat area just above the lateral orbital edges in trigonocephaly. Periosteum, subcutis, and cutis are closed in separate layers using resorbable sutures, Steristrips™ (3M™, Diegem, Belgium) included. A small compressive head bandage is used for 24 h. Facial and periorbital swelling is usually very mild.

Anterior plagiocephaly/brachycephaly

Patients are positioned in the supine position with the head contralaterally rotated in plagiocephaly cases or neutral position in brachycephaly cases. One (or two) curvilinear skin incision of approximately 3 cm wide is placed just behind the hairline over the synostotic suture(s). Depending on the hairline and the specific curvature of the forehead, we sometimes use a small zigzag incision (Harry Potter incision) to allow better skin retraction [Figure 4]. From this skin incision, an osteoclastic craniectomy towards the anterior fontanel is performed using the high-speed drill and rongeurs after dissection and elevation of the periosteum [Video 2]. The length of this craniectomy can vary in case a part of the suture is still open and patent. After this, FloSeal® Matrix Hemostatic Sealant is administered for hemostasis. Then, the endoscope is introduced and dura dissection from the overlying bone is performed up to the pterion. The perfect visualization of the dura and operative field by the endoscope in conjunct with a parallel positioned aspirator to clear any blood allows a safe dissection of the dura, without any problems with the middle meningeal artery branches. Once the dura dissection is completed, the periosteum is dissected and lifted from the suture. The synostotic suture is then removed with a width of 1–2 cm. At the pterion, some

thick, cancellous bone can be encountered which may be responsible for some venous bleeding. This can easily be controlled by using FloSeal® Matrix Hemostatic Sealant, and Ostene® bone wax. Periosteum, subcutis, and cutis are closed in separate layers using resorbable sutures, Steristrips™ (3M™, Diegem, Belgium) included. A small compressive head bandage is used for 24 h.

Multisutural and syndromic craniosynostosis

Although our experience is small for multisutural, nonsyndromic cases, we adhere to the same rationale for performing ECAS in these cases as for monosutural synostosis. Jimenez and Barone have shown that nonsyndromic multisutural craniosynostosis can be treated successfully with excellent results and reversal of the deformities.^[21] Therefore, we use the same timing and technique as in monosutural synostosis cases, including helmet therapy. Positioning depends on the affected sutures and is aimed at including all affected sutures within one sterile operative field. In general, all affected sutures are treated with suturectomy through one skin incision for every suture.

Recently, Jimenez and Barone reported on the endoscopic-assisted bilateral strip craniectomy of the coronal suture in an infant with Apert syndrome followed by helmet therapy.^[15] Based on their experience and short-term follow-up, they stated that early endoscopic-assisted surgery may provide an alternate and safe surgical option to treat complex syndromic craniosynostosis, although long-term results are needed to evaluate this.

In our centre, we treated three Apert and two Muenke syndrome cases with EACS. However, we did so with a different goal than Jimenez *et al.* In syndromic craniosynostosis, we want to try to halt the progressive deformity, prevent intracranial hypertension, and simplify reconstructive surgery at a later stage by performing EACS in a very early stage (4–8 weeks of age) but without helmet molding therapy. This is a very easy and simple surgery with very low morbidity, but to our mind, it is not meant to be a replacement for conventional surgical techniques. It is rather a supplement treatment

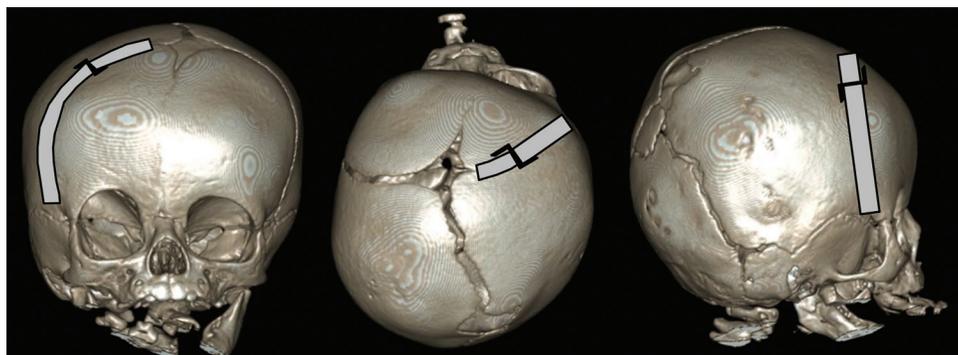


Figure 4: 3D scan showing extent of craniectomy in plagiocephaly. Thick black line indicates skin incision, grey area depicts craniectomy size

to reduce the burden of the syndrome on the infant until definitive reconstructive surgery can be performed at a later age. Some groups have started to perform posterior cranial vault expansion in a minimally invasive manner to achieve the same goal while waiting for the right time to perform definitive cranial vault reconstruction.^[2]

Helmet therapy

To our mind, the success of EACS depends heavily on the cranial molding therapy. The helmet design, subsequent modifications, and compliance to the helmet therapy are all critical to the success of this procedure. The helmet has the ability to modify the calvarial growth pattern, and hence, the direction of growth in three dimensions. By controlling growth in most areas, the helmet focuses most of cranial growth in the areas where it is needed. By guiding the cranial growth in three dimensions, the fast developing and growing brain can act as a very effective internal distractor once suturectomy is performed. Without this guidance, e.g., due to lack of fit or noncompliance, cranial expansion occurs equally in all directions and the obtained correction after suturectomy remains incomplete.

One week after the surgery, a plaster imprint of the skull is taken, which serves as an initial template for the fabrication of the custom-made helmet and helmet therapy starts within 2 weeks after surgery. The helmet is designed to contact all areas of the cranium except where growth is desirable. Because the helmet needs to be worn 23 h daily, a perfect fit of the helmet is of paramount importance to prevent slippage or the development of pressure ulceration areas or other skin problems. Frequent follow-up by a dedicated orthotist and the craniofacial team, especially at the early stage of the therapy, ensures a perfect fit and allows for patient-specific adjustments in reaction to actual skull growth in three dimensions. This can be easily done by thermoplastic procedures until skull growth requires a new helmet. As the fastest increase in cranial volume occurs during the first two years of life, we aim for continuing helmet therapy until the age of 1–1.5 years or when normocephaly has been reached. In our series, the helmet was worn for 10 months on average.^[6] Children needed 1 or 2 helmets in the beginning of our experience. As our procedure and the importance of early referral to our centre was slowly adopted by the healthcare system, we were able to shift the timing of the surgery more towards the age of 3 months. Being treated earlier, most children need now 2 to 3 helmets during treatment. As our experience with this procedure grew, we adjusted the design of the helmet in close collaboration with the orthotist. At this stage, we use two different types of helmet according to the involved suture. For scaphocephaly, a recently developed very light, one-piece resin helmet is used [Figure 5]. This helmet has a thickness of only 6 mm and reaches very low at the back of the head and encloses the entire forehead.



Figure 5: Helmets used for orthotic treatment. Left: 2-piece thermoplastic helmet used for trigonocephaly/anterior plagiocephaly. Right: one-piece resin helmet used for scaphocephaly

This allows for a perfect fit, no slippage, and no need for a chin closure. It has limited ability for thermoplastic adjustments and is somewhat stiffer, exerting a bigger force in anterior-posterior direction. With this helmet, we notice that occipital rounding of the head as well as frontal bossing tends to correct faster. However, this needs to be verified in the future with increasing patient numbers. For trigonocephaly, brachycephaly, and plagiocephaly, a two-piece plastic helmet is used [Figure 5]. This helmet is slightly thicker, 8 mm, and allows the correction of the forehead as needed in these cases. Again, it reaches very low at the back of the head as well as at the nasion, without obstructing vision. This allows for a perfect fit, no slippage, and no need for a chin closure. This helmet is made of a thermoplastic material, allowing for more easy adjustments by heating. Especially in plagiocephalic cases, where asymmetry needs to be addressed, this allows for frequent adjustments according to the local skull growth, when the affected side is changing faster than the general growth of the skull. This resolves the need for constructing a new helmet for a local change, while still being able to guide local skull growth.

DISCUSSION

Recent reports focus on the embryological formation and premature closure of sutures as being the main pathogenetic cause for craniosynostosis to occur.^[27,33] Thus, it starts with a prematurely closed suture and subsequently the resultant cranial deformity is mostly the result of compensatory overgrowth at adjacent sutures, as Delashaw showed in 1989.^[5] This is a strong argument to try to perform surgery as soon as possible to interact and halt the further developing cranial deformity. To our mind, this is where technological advances make the difference; by using endoscopic techniques, the morbidity and mortality of surgery has dramatically dropped,

allowing surgery in very young children. Technology can also overcome the shortcomings of simple suturectomy reported earlier. Based on Moss' functional matrix theory, the brain can be used as a perfect internal distractor once suturectomy is performed, but it needs guidance.^[28]

By using an orthotic molding helmet, the distractive forces of the growing brain can be guided towards the preferable growing vectors in three planes. We think that EACS with helmet therapy is the next logical step in the evolution of surgical techniques for craniosynostosis as it results from the combination of new insights into the pathogenetic mechanisms at play, together with the development of new technologies.

After having performed more than 140 cases, including all types of monosutural as well as complex nonsyndromic multisutural and some syndromic cases, we consider this technique as a very safe and valuable tool in the broad range of treatment possibilities for craniosynostosis, with satisfying results [Figures 6-8]. With our current experience, we actively advise this treatment to any craniosynostosis patient under the age of 4 months, but for patients aged 4–5 months with moderate-to-severe craniosynostosis (especially plagiocephaly and trigonocephaly), we inform parents that this treatment may not be sufficient and cranial vault reconstruction techniques may have to be performed at a later stage. Up to now, this was needed three times, of which one was for cosmetic reasons only. In this particular case of multisutural craniosynostosis involving the left coronal and sagittal suture, left plagiocephaly persisted after EACS and helmet therapy.^[6]

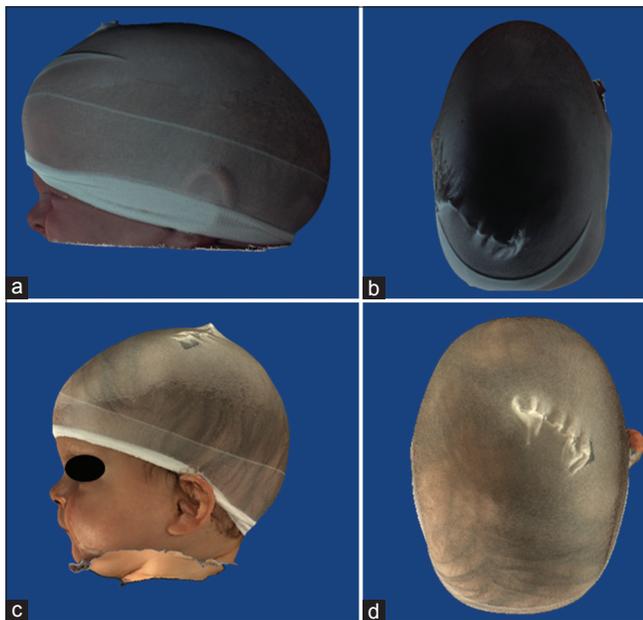


Figure 6: (a and b) pre operative 3D fotogrammetry of a scaphocephalic patient. (c and d) 11 months postoperative 3D fotogrammetry of same patient. Frontal bossing has declined, occipital pointing is resolved, mid-parietal breadth normalized

However, as experience grows and long-term follow-up data starts to be available in the literature, we think it is mandatory to evaluate the results of this technique by comparing them not only to historical data but also to other current techniques including both minimally invasive spring-assisted techniques as classical “open” techniques. Indeed, also classic “open” cranial vault reconstruction techniques have undergone some recent adjustments to reduce surgical time, blood loss, and morbidity. It is mandatory to try to evaluate whether EACS is suitable for all craniosynostosis cases, both nonsyndromic and syndromic, or only for selected subgroups.

The result of the EACS treatment depends heavily on the helmet therapy. This essential part of the treatment is often considered a major drawback of this treatment, some even call it a “complication” of this treatment (personal communication at VI World congress of Neuroendoscopy, Mumbai, 2013). In our series, helmet therapy was continued for a mean of 10 months (8–12 months). By using custom-made, very light helmets, the compliance rate of helmet therapy is very high and we never noticed pressure ulcers or major complications. In few cases, some eczema or dry skin developed, which resolved once helmet therapy was stopped. Up to now, we never had to stop helmet therapy because of intolerance.



Figure 7: (a and b) pre operative 3D fotogrammetry of a trigonocephalic patient. (c and d) one year postoperative 3D fotogrammetry of same patient. The width of the forehead is already increased, there is still some backslanting of the lateral brow

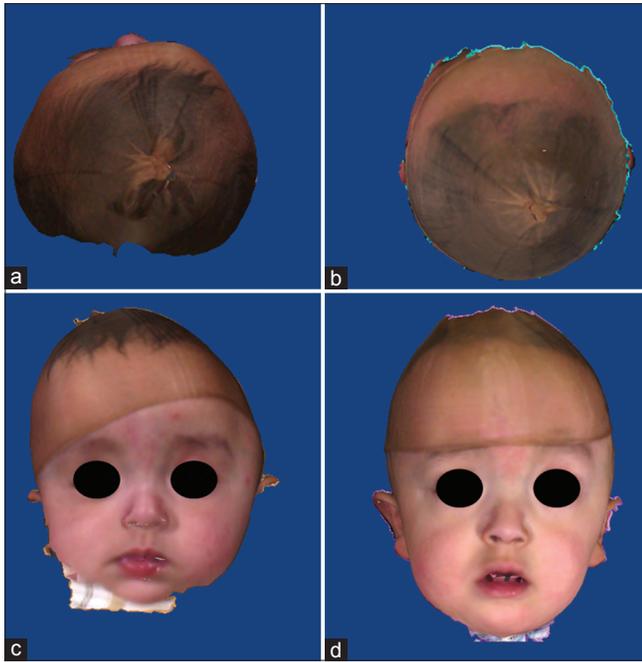


Figure 8 : (a and b) pre operative 3D fotogrammetry of a patient with synostosis of left coronal and bilateral lambdoid sutures. (c and d) 12 months postoperative 3D fotogrammetry of same patient. The cranial axis has almost completely aligned with the facial axis and the shape of the forehead is almost symmetrical, with perfect rounding of the occipital area

Overall, the “burden” of the helmet therapy, as reported by parents, seems to be very low. We tried to generate objective data on the burden of helmet therapy by sending an online questionnaire to all parents. The questions in the questionnaire covered all areas of the impact and were asked objectively. This does not rule out all bias, e.g., it can be that parents choosing a type of procedure are less likely to report that they made a mistake in choosing. Although only roughly one-third of all parents responded, results are mainly in favor for the helmet therapy. Almost everyone would choose again the EACS with helmet therapy and all respondents would advise others to choose this treatment.^[6] This is in agreement with reports by other centers treating large numbers of patients with EACS.^[3,17,18,35] However, the so called “burden” of the helmet therapy remains one of the main arguments to discard this treatment by those who have no experience with this treatment and are ill-informed.

When looking at the key points of this treatment, we still see room for future improvements:

- Patients need to be treated as early as possible, preferably just before or at the age of 3 months to get satisfying results. Therefore, early referral to the neurosurgeon is of paramount importance. Information and education of general practitioners, pediatricians, and paramedic professionals working with children with “abnormal” head shapes (physiotherapists, manual therapists, etc.) should

be actively performed to raise awareness and enlarge the therapeutic time frame through early referrals.

- Although custom available surgical instruments are sufficient to perform EACS safely and successfully, development of dedicated instruments for this type of surgery can improve efficiency and reduce surgical time and blood loss even further. Especially the development of a new type of endoscopic shaft, small dedicated instruments for hemostasis, and a new design of a small craniotome that can be easily used below the skin under endoscopic guidance would improve surgical technique.^[39]

- Helmet design can be further refined by using 3D CAD techniques. This could allow to construct the perfect molding helmet taking into account the actual skull compared to a reference “normal” skull to define the areas and extent of desired growth and/or restriction of growth. This would make the helmet therapy more reliable and predictable, with easier, planned, adaptations. This technique is already available but at the moment is more expensive than handmade custom helmets.

Last but not least, EACS may be combined with other surgical techniques as well. Spring expansion, internal and external distraction, and orbitofrontal advancement may all be combined with EACS, wherein the combination of two techniques allows further improvement of the result.

To our mind, the biggest challenge for the coming decades in the field of craniosynostosis surgery is trying to define which surgical technique or combination of techniques – open, endoscopic, spring-assisted – yields the best results in terms of satisfying cosmetic and functional results, with the lowest morbidity, mortality and cost, for certain set parameters such as craniosynostosis subtype, degree of severity, age at presentation, gender, and genetic background.

However, the field of craniosynostosis surgery is currently limited by the lack of objective data by which to interpret and compare results.^[7] Comparison is often performed on subjective basis and biased by personal experience, teaching schools, training, and dogmas, although there are increasing efforts to develop objective tools to evaluate cosmetic outcomes. We agree fully with Hankinson *et al.* who stated “Until a satisfactory craniometric method or group of methods is established, it will be difficult to meaningfully compare the outcomes of the myriad operative techniques currently available for the treatment of single suture craniosynostosis,” and by extension multisuture craniosynostosis.^[11]

CONCLUSION

We think EACS with molding helmet therapy offers an excellent alternative to traditional open approaches and should be considered for children diagnosed with

nonsyndromic craniosynostosis prior to 3 months of age. Based on our very limited experience, we think it might also be a meaningful add-on therapy for syndromic cases to relieve the burden of the syndrome on the infant until definitive reconstructive surgery can be performed at a later age. In our experience, the helmet molding therapy is essential for reaching good results. This therapy is well tolerated by the children and parents alike without any major complications or concerns.

Financial support and sponsorship

Nil.

Conflicts of interest

None of the authors have any conflict of interest with publication of the manuscript or an institution or product that is mentioned in the manuscript and/or is important to the outcome of the study presented.

REFERENCES

- Anderson FM, Johnson FL. Craniosynostosis; a modification in surgical treatment. *Surgery* 1956;40:961-70.
- Arnaud E, Marchac A, Jebblou Y, Renier D, Di Rocco F. Spring-assisted posterior skull expansion without osteotomies. *Childs Nerv Syst* 2012;28:1545-9.
- Berry-Candelario J, Ridgway EB, Grondin RT, Rogers GF, Proctor MR. Endoscope-assisted strip craniectomy and postoperative helmet therapy for treatment of craniosynostosis. *Neurosurg Focus* 2011;31:E5.
- Chan JW, Stewart CL, Stalder MW, St. Hilaire H, McBride L, Moses MH. Endoscope-assisted versus open repair of craniosynostosis: A comparison of perioperative cost and risk. *J Craniofac Surg* 2013;24:170-4.
- Delashaw JB, Persing JA, Broaddus WC, Jane JA. Cranial vault growth in craniosynostosis. *J Neurosurg* 1989;70:159-65.
- Delye HHK, Arts S, Borstlap WA, Blok LM, Driessen JJ, Meulstee JW, et al. Endoscopically assisted craniosynostosis surgery (EACS): The craniofacial team Nijmegen experience. *J Craniofac Surg* 2016;44:1029-36.
- Delye H, Clijmans T, Mommaerts MY, Vandersloten J, Goffin J. Creating a normative database of age-specific 3D geometrical data, bone density and bone thickness of the developing skull: A pilot study. *J Neurosurg Pediatr* 2015;16:687-702.
- Faber HK, Towne EB. Early craniectomy as a preventive measure in oxycephaly and allied conditions: With special reference to the prevention of blindness. *Am J Med Sci* 1927;173:701-11.
- Faber HK, Towne EB. Early operation in premature cranial synostosis for the prevention of blindness and other sequelae. Five case reports with follow-up. *J Pediatr* 1943;22:286-307.
- Greene CS Jr, Winston KR. Treatment of scaphocephaly with sagittal craniectomy and biparietal morcellation. *Neurosurgery* 1988;23:196-202.
- Hankinson TC, Fontana EJ, Anderson RCE, Feldstein NA. Surgical treatment of single-suture craniosynostosis: An argument for quantitative methods to evaluate cosmetic outcomes. *J Neurosurg Pediatr* 2010;6:193-7.
- Ingraham FD, Alexander E Jr, Matson DD. Clinical studies in craniosynostosis analysis of 50 cases and description of a method of surgical treatment. *Surgery* 1948;24:518-41.
- Jacobi A. Non nocere. *Med Rec* 1894;45:609-18.
- Jane JA, Edgerton MT, Futrell JW, Park TS. Immediate correction of sagittal synostosis. *J Neurosurg* 1978;49:705-10.
- Jimenez DF, Barone CM. Bilateral endoscopic craniectomies in the treatment of an infant with Apert Syndrome. *J Neurosurg Pediatr* 2012;10:310-4.
- Jimenez DF, Barone CM. Early treatment of coronal synostosis with endoscopy-assisted craniectomy and postoperative orthosis therapy: 16-year experience. *J Neurosurg Pediatr* 2013;12:207-19.
- Jimenez DF, Barone CM. Endoscopic technique for coronal synostosis. *Childs Nerv Syst* 2012;28:1429-32.
- Jimenez DF, Barone CM. Endoscopic technique for sagittal synostosis. *Childs Nerv Syst* 2012;28:1333-9.
- Jimenez DF, Barone CM. Endoscopic craniectomy for early surgical correction of sagittal craniosynostosis. *J Neurosurg* 1998;88:77-81.
- Jimenez DF, Barone CM. Endoscopy-assisted wide-vertex craniectomy, "barrel-stave" osteotomies, and postoperative helmet molding therapy in the early management of sagittal suture craniosynostosis. *Neurosurg Focus* 2000;9:e2.
- Jimenez DF, Barone CM. Multiple-suture nonsyndromic craniosynostosis: Early and effective management using endoscopic techniques. Clinical article. *J Neurosurg Pediatr* 2010;5:223-31.
- Jimenez DF, Barone M, Cartwright CC, Baker L. Early Management of Craniosynostosis Using Endoscopic-Assisted Strip Craniectomies and Cranial Orthotic Molding Therapy. *Pediatrics* 2002;110:97.
- Lane LC. Pioneer craniectomy for relief of mental imbecility due to premature sutural closure and microcephalus. *JAMA* 1892;18:49-50.
- Lannelongue M. De la craniectomie dans la microcéphalie. *Compt Rend Seances Acad Sci* 1890;50:1382-5.
- Lauritzen C, Sugawara Y, Kocabalkan O, Olsson R. Spring mediated dynamic craniofacial reshaping. Case report. *Scand J Plast Reconstr Surg Hand Surg* 1998;32:331-8.
- Mehta VA, Bettegowda C, Jallo GI, Ahn ES. The evolution of surgical management for craniosynostosis. *Neurosurg Focus* 2010;29:E5.
- Morriss-Kay GM, Wilkie AOM. Growth of the normal skull vault and its alteration in craniosynostosis: Insights from human genetics and experimental studies. *J Anat* 2005;207:637-53.
- Moss ML. The pathogenesis of premature cranial synostosis in man. *Acta Anat (Basel)* 1959;37:351-70.
- Okada H, Gosain AK. Current approaches to management of nonsyndromic craniosynostosis. *Curr Opin Otolaryngol Head Neck Surg* 2012;20:310-7.
- Proctor MR. Endoscopic cranial suture release for the treatment of craniosynostosis-is it the future? *J Craniofac Surg* 2012;23:225-8.
- Rivero-Garcia M, Marquez-Rivas J, Rueda-Torres AB, Ollero-Ortiz A. Early endoscopy-assisted treatment of multiple-suture craniosynostosis. *Childs Nerv Syst* 2012;28:427-31.
- Sanger C, David L, Argenta L. Latest trends in minimally invasive synostosis surgery: A review. *Curr Opin Otolaryngol Head Neck Surg* 2014;22:316-21.
- Senarath-Yapa K, Chung MT, McArdle A, Wong VW, Quarto N, Longaker MT, et al. Craniosynostosis, Molecular pathways and future pharmacologic therapy. *Organogenesis* 2012;8:103-13.
- Sgouros S. Skull vault growth in craniosynostosis. *Childs Nerv Syst* 2005;21:861-70.
- Shah MN, Kane AA, Petersen JD, Woo AS, Naidoo AD, Smyth MD. Endoscopically assisted versus open repair of sagittal craniosynostosis: The St. Louis Children's Hospital experience. *J Neurosurg Pediatr* 2011;8:165-70.
- Shillito Jr, Matson DD. Craniosynostosis: A review of 519 surgical patients. *Pediatrics* 1968;41:829-53.
- Simmons DR, Peyton WT. Premature closure of the cranial sutures. *J Pediatr* 1947;31:528-47.
- Tessier P. The definitive plastic surgical treatment of the severe facial deformities of craniofacial dysostosis. Crouzon's and Apert's diseases. *Plast Reconstr Surg* 1971;48:419-42.
- Verhamme L. Craniosynostosis surgery: Development of a flexible, angled craniotome. MSc. Thesis, Faculty of Mechanical, Maritime and Materials Engineering, Delft University of Technology, Delft, Netherlands; 2009.
- Virchow R. Über den Cretinismus, namentlich in Franken, und über pathologische Schädelformen. *Verh Phys Med Gesell Würzburg* 1851;2:230-71.