

# Diagnosis and treatment of craniosynostosis: Vilnius team experience

Linus Zaleckas<sup>3</sup>,

Arijanda Neverauskienė<sup>1,2</sup>,

Vidūnas Daugelavičius<sup>1,4</sup>,

Dominika Šidlovskaitė-Baltakė<sup>4</sup>,

Ramūnas Raugas<sup>1</sup>,

Berta Vištartaitė<sup>1</sup>,

Evelina Balčiūnaitė<sup>1</sup>

<sup>1</sup> Faculty of Medicine,  
Vilnius University, Lithuania

<sup>2</sup> Centre of Radiology  
and Nuclear Medicine,  
Children's Hospital,  
Affiliate of Vilnius University  
Hospital Santariškių Klinikos

<sup>3</sup> Centre of Oral and  
Maxillofacial Surgery,  
Institute of Odontology,  
Faculty of Medicine,  
Vilnius University

<sup>4</sup> Centre of Paediatrics Intensive  
Care and Anaesthesiology,  
Children's Hospital,  
Affiliate of Vilnius University  
Hospital Santariškių Klinikos

**Background.** The aim of the study was to review the methods of diagnosis of craniosynostosis and to analyze Vilnius (Lithuania) team experience of surgical treatment, surgical methods, aspects of anesthesia for patients with craniosynostosis and to present early results of surgical treatment.

**Materials and methods.** A retrospective review of all patients with various types of craniosynostosis treated surgically during the period from 1 January 2009 to 31 December 2014 was performed. The following data were analyzed: age, type of deformity, surgical technique, surgical time, methods and course of anesthesia, intra- and postoperative complications, parents' satisfaction, head form.

**Results.** 24 patients were treated. The mean patient's age at the time of surgery was  $13.47 \pm 8.2$  months (min 7.3, max 46.5). Eliminating 3 patients whose age at the time of surgery was over 2 years (24.5, 29 and 46 months, respectively), the mean age of other 21 patients was  $10.63 \pm 1.77$  (min 7.3, max 14.1) months. There were 9 cases of isolated trigonocephaly (37.5%), 7 cases of isolated scaphocephaly (29.2%), 7 cases of isolated anterior plagiocephaly (29.2%) and 1 case of posterior plagiocephaly combined with scaphocephaly (4.17%). All craniosynostoses were diagnosed clinically and diagnosis was confirmed with computed tomographic scanning. The median duration of surgery was  $336.47 \pm 59.63$  minutes (min 308.13, max 364.82). The median stay in the intensive care unit was  $2.53 \pm 1.28$  days (min. 1.92, max 3.14). In all cases rigid osteosynthesis was performed. 2 children were diagnosed with syndromic craniofacial abnormalities. In 23 (95.83%) cases an intraoperative or postoperative blood transfusion was required. In 24 treated patients there was no mortality. During the early and late postoperative period no infections, CSF leakage and dural tears were observed. No neurological impairments or any signs of neurological deficits were observed by any of the treated patients. In all of cases parents were satisfied with their children's changed head shape and aesthetic results.

**Conclusions.** Cranioplasties for correction of craniosynostosis give good aesthetic results and this is a safe method, which helps to correct the head shape as well as improves the social adaptation of patients.

**Key words:** craniosynostosis, tomography, X-Ray Computed, craniofacial deformity, craniofacial surgery, imaging

Correspondence to: Arijanda Neverauskienė, Children's Hospital, Affiliate of Vilnius University Hospital Santariškių Klinikos, Santariškių St. 7, LT-08406 Vilnius, Lithuania. E-mail: arijandanever@gmail.com

## INTRODUCTION

Craniosynostosis is the premature closure of one or more of the cranial sutures leading to an abnormal skull shape (1). It occurs within the population with a frequency of 1:2,000 to 1:4,000 live births (2, 3). Abnormal head shapes are therefore produced because of the restricted growth, which occurs perpendicular to the fused sutures and to the compensatory growth in the skull's unfused bony plates (4). Craniosynostosis results in deformed calvaria at births (5). This deformation usually increases during the first months after births. It is not only esthetic problem – the deformed skull also affects brain growth in to the affected direction and in the case of uncorrected condition often leads to decreased mental ability.

The indications for treatment of craniosynostosis skull deformity relate primarily to correcting skull deformity and improving mental function (6).

Although the craniosynostosis is one of the most frequent craniofacial malformations (7), it continues to be a diagnostic and/as well therapeutic challenge which requires the cooperation of various medical specialties – general practitioners, pediatricians, neuro- and maxillofacial surgeons, anesthesiologists, radiologists and many other specialists in diagnosis managing and treatment of this disorder.

The optimal time for operative treatment of craniosynostosis still remains a point of discussion although some surgeons tend to operate these patients earlier than 6 months of age, some tend to operate these patients two or three months later. One of the most important factors that can support the optimal treatment is the early recognition/identification of craniosynostotic pathology and referring them to the craniofacial surgeons.

The first modern cranioplasty for correction of craniosynostosis in Lithuania was performed on 18 February 2009 (8). Until this time no craniofacial surgery correcting the fronto-orbital segment was performed in Lithuania.

Until 2009 craniosynostotic surgery was underdeveloped in Lithuania. Only simple decompressing linear osteotomies performed by neurosurgeons were made. This was caused by many factors – lack of craniomaxillofacial surgeons interesting in this field, lack of cooperation between

the various medical specialties, lack of modern surgical and anesthesiological equipment. One of the most important factors was also poor or insufficient physician's knowledge about craniosynostosis. This caused the delayed referring of these patients to the craniofacial specialist. Due to lack of knowledge it was believed that any bigger/modern craniofacial surgery is too aggressive and too dangerous for craniosynostotic children. Patients with craniosynostotic pathology, especially involving the fronto-orbital region, became almost without any professional help/assistance.

The aim of the study was to review the methods of diagnosis of craniosynostosis and to analyze the Vilnius (Lithuania) team experience of surgical treatment, surgical methods, aspects of anesthesia for patients with craniosynostosis and to present early results of surgical treatment.

## MATERIALS AND METHODS

The medical records of patients with craniosynostosis treated surgically during the period from 1 January 2009 to 31 December 2014 were reviewed. During this five-year period 24 patients with various types of craniosynostosis were treated.

A retrospective review of all medical records of patients treated due to craniosynostosis and head deformity was performed. The following data were analyzed: age, type of deformity, surgical technique, surgical time, methods and course of anesthesia, intra- and postoperative complications, parent's satisfaction, head form.

Diagnosis of craniosynostosis was confirmed after a history, general examination and computed tomographic scanning.

Patients were also investigated during regular follow-up visits at 2, 4 weeks, 3 and 6 months after surgery and later yearly.

## RESULTS

During the period from 1 January 2009 to 31 December 2014, twenty four cranioplasties were performed. The amount of surgeries varied from year to year. The first cranioplasty correcting anterior plagiocephaly was performed on 18 February 2009 (8). During the year 2010 three and in the year 2011 four open cranioplastic surgeries were

done. The number of surgeries was started to grow in the year 2012 and during this year 8 cranioplasties were performed, but in the next year (2013) only 1 cranioplastic surgery was done. In 2014 again 7 patients correcting their craniosynostotic condition were treated. The minimal follow-up period was 6 months, the maximal one was 6 years.

The surgical pathology included various types of head deformations. There were 9 cases of isolated trigonocephaly (37.5%), 7 cases of isolated scaphocephaly (29.2%), 7 cases of isolated anterior plagiocephaly (29.2%) and 1 case of posterior plagiocephaly combined with scaphocephaly (4.17%).

The mean patient's age at the time of surgery was  $13.47 \pm 8.2$  months (min 7.3, max 46.5). Eliminating 3 patients whose age at the time of surgery was over 2 years (24.5, 29 and 46 months, respectively), the mean age of other 21 patients was  $10.63 \pm 1.77$  (min 7.3, max 14.1) months.

The median duration of surgery was  $336.47 \pm 59.63$  minutes (min 308.13, max 364.82). The last surgeries were performed in shorter time than the first surgeries but it was statistically insignificant ( $p > 0.05$ ).

The medium stay in the intensive care unit was  $2.53 \pm 1.28$  days (min 1.92, max 3.14).

### **Surgical technique and anesthesia**

All patients were assessed as ASA 1–2. The patients were premedicated with midazolam 0.25 mg. Normothermia was facilitated by a forced air warmer and an intravenous fluid warmer. The operating room temperature was maintained at 25 °C. The patients were anesthetized with an inhalation induction with Sevoflurane in oxygen and an air mixture by a mask. After intravenous access was secured, a bolus of fentanyl ( $1\text{--}2 \mu\text{g}\cdot\text{kg}^{-1}$ ) and a muscle relaxant (Rocuronium  $0.6 \text{ mg}\cdot\text{kg}^{-1}$ ) was administered to facilitate tracheal intubation. After the tracheal intubation the central venous line was inserted under the guidance of ultrasound. Anesthesia was maintained with Sevoflurane in an air/oxygen mixture with co-administration of fentanyl infusion ( $1\text{--}3 \mu\text{g}\cdot\text{kg}^{-1}\cdot\text{h}^{-1}$ ). During anesthesia, monitoring of ECG,  $\text{SpO}_2$ , anaesthetic agent, capnography, NIBP, temperature and urine output were performed. Additionally Masimo Rainbow Pulse CO-Oximetry ( $\text{SpO}_2$ ; SpHb; SpOC; SpCO; SpMet: PR; PI; PVI) and NIRS were used in the

last three years. Intraoperative blood loss was estimated by the anesthesia and surgical teams.

Intraoperative intravenous fluids were acetate Ringer, normal saline (0.9% sodium chloride) and colloid (Voluven 6%). Intravenous fluids were administered to maintain an adequate intravascular volume based on the heart rate, systolic pressure variation and urine output. Allogenic blood transfusion with packed red blood cells was administered for hemoglobin  $<70 \text{ g/l}$  or  $>70 \text{ g/l}$  at hemodynamic instability and ongoing blood loss. In the last four cases Mannitol ( $0.5\text{--}1 \text{ g/kg}$ ) was used to reduce intracranial hypertension and cerebral edema on request of surgeons. Additionally, for these patients a tranexamic acid (TXA) bolus ( $10 \text{ mg}\cdot\text{kg}^{-1}$ ) was administered intravenously about 30 minutes before surgery to reduce intraoperative blood loss. We started another TXA dose of  $10 \text{ mg}\cdot\text{kg}^{-1}$  IV infusions for 4 hours ( $2.5 \text{ mg}\cdot\text{kg}^{-1}\cdot\text{h}^{-1}$ ) during surgery after patient's positioning on the operation table.

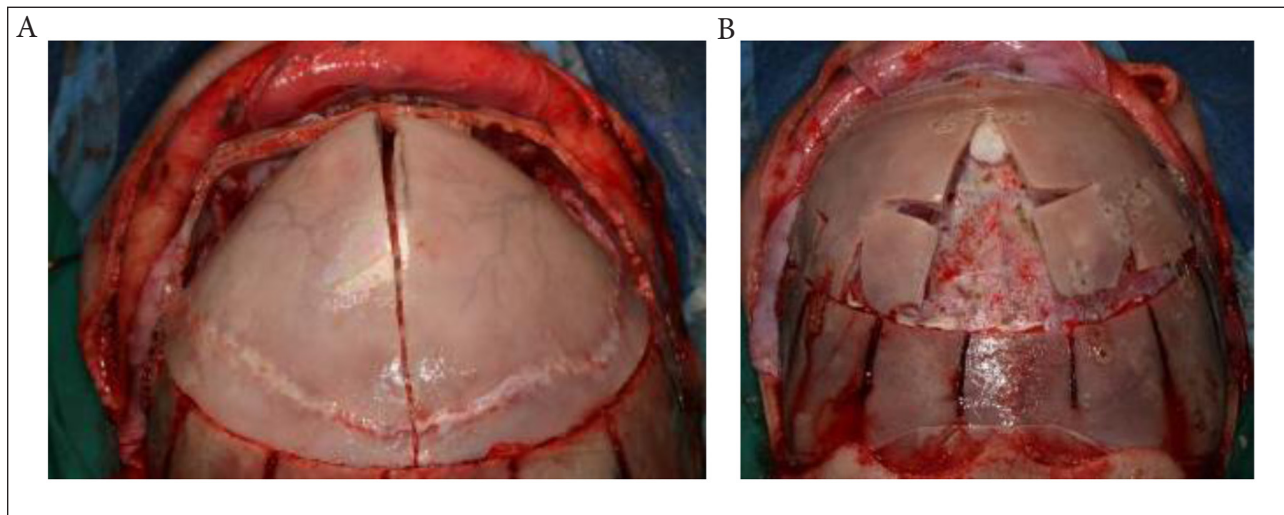
In the Pediatric Intensive Care Unit the quantity and tonicity of postoperative intravenous fluids (acetate Ringer, 0.9% sodium chloride, 5% or 10% glucose solution), electrolyte correction and allogenic blood transfusion (packed red blood cells) were administered under the direction of the pediatric intensive care specialists. Crystalloids and glucose solutions supplemented with NaCl 10% (53%) and KCl (71%) were used.

Postoperatively three children received red blood cells transfusion (12.5%) additionally and one (4.17%) received Albumin 20%. Postoperative analgesia was provided with morphine.

In all cases of anterior plagiocephaly or trigonocephaly corrections ( $n = 11$ ), in the supine position the remodeling of the anterior cranial vault and frontoorbital complex and the reconstruction of orbits were applied.

After bicoronal waved or zig-zag incision, the skin and galeal layer was raised. After that the periosteal layer was raised creating a periosteal flap. The frontal bone including the bicoronal suture is removed in one piece followed by deivision and elevation of the supraorbital segment.

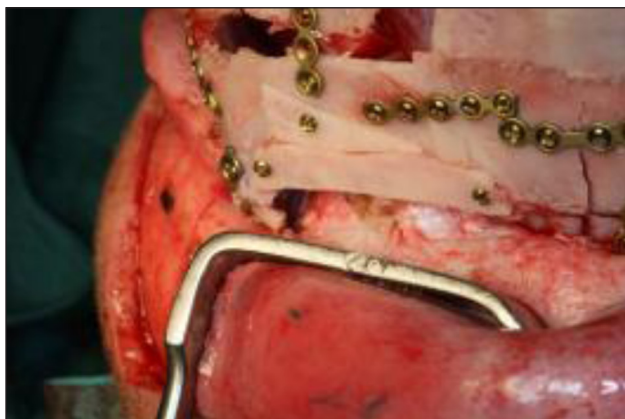
In the case of trigonocephaly the supraorbital bandeau was reshaped creating eyebrow prominences and fixed into the new position increasing the interorbital distance and increasing the fronto-temporal angle (Fig. 1).



**Fig. 1.** A. Reshaped and to the new position fixed supraorbital bandeou. The frontal bone was cut in the midline and placed into the old position for comparison. B. Remodeled frontal bone fitted to the fronto-orbital bar. Linear osteotomies of the parietal bone helps to enlarge the cranial volume in the anterior part of the cranium

The frontal bone is cut in the midline and divided into two pieces and remodeled to fit to the new shape of the supraorbital bar leaving both coronal sutures in the parallel position to the supraorbital osteotomy line (9). The fixation of bone fragments was performed using rigid osteosynthesis and some places of bone fragments were fixed using resorbable sutures (2-0 and 3-0 Vicryl).

Correcting the plagiocephalic condition various methods of bone remodelling were applied – reshaping of the frontoorbital bandeau (in all cases), dividing of the frontal bone into two pieces, reshaping of the frontal bone, bone augmentation creating eyebrow prominence in the affected site (Fig. 2).



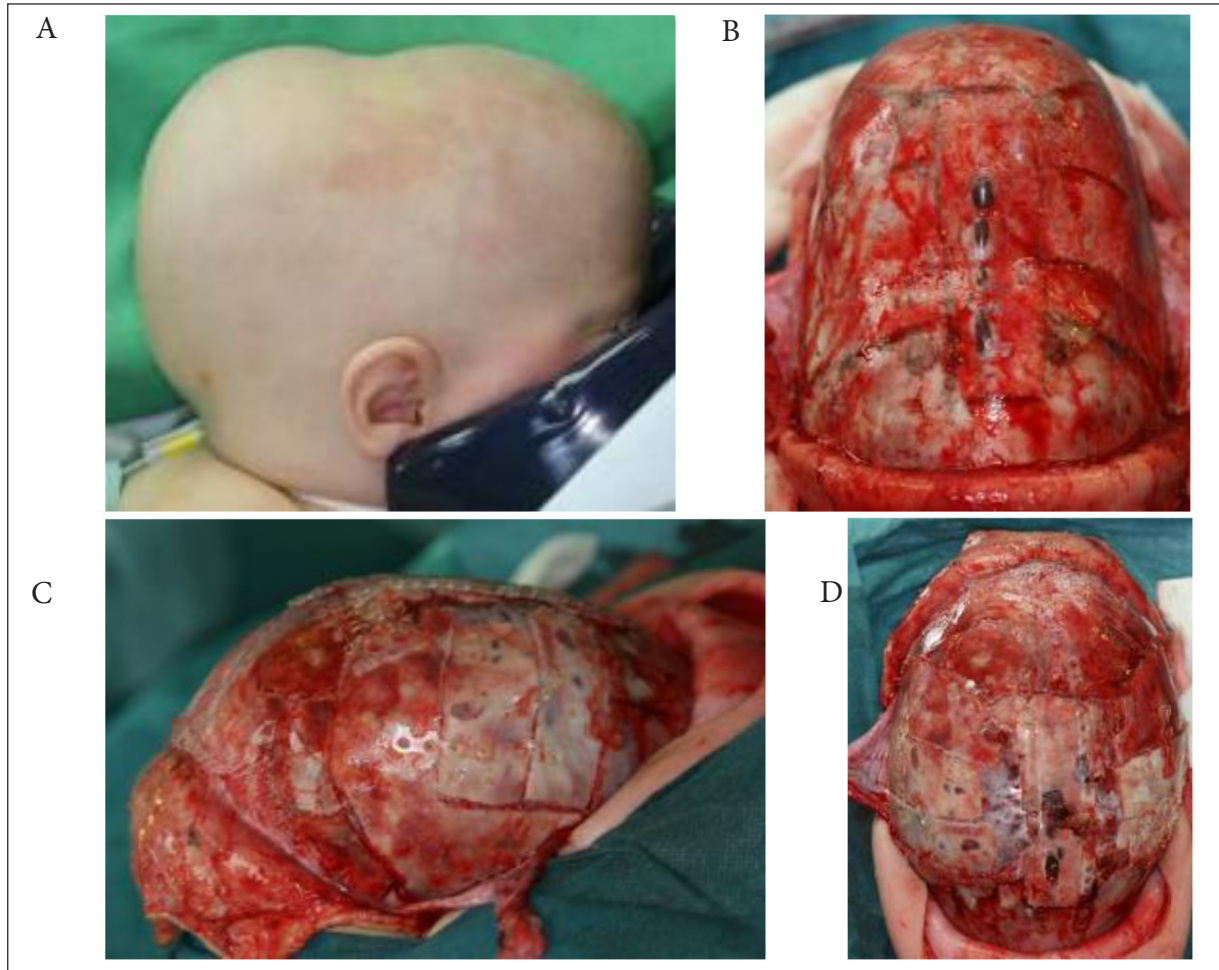
**Fig. 2.** Augmented eyebrow prominence. Bone fragments are fixed with titanium microplates

The corrections of scaphocephalies were performed in the prone or modified prone position. Various surgical techniques were used due to different cranial shape and patient's age (Figs. 3, 4).

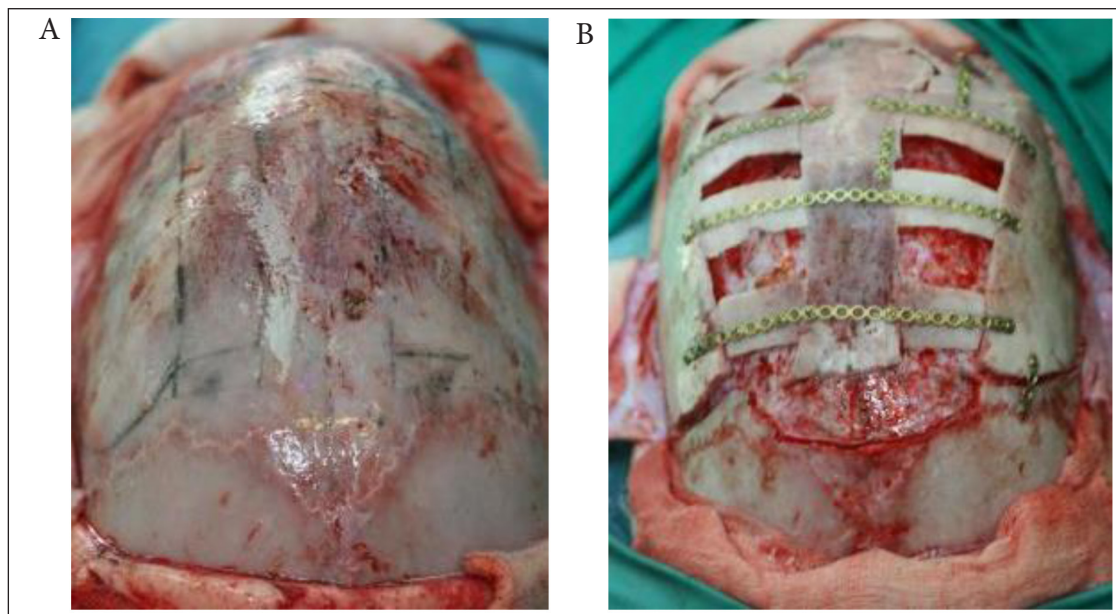
In all cases rigid osteosynthesis was performed: in 11 cases titanium microplates were used, in another 13 cases resorbable plates (12 cases – SonicWeld Rx®, KLS Martin and 1 case – lactosorb Lorenz Biomet) were applied. When osteosynthesis of reshaped cranial bones was performed with titanium microplates (titanium microplates, produced by Stryker Leibinger GmbH & Co.), the second stage surgery for elimination of plates was required. The period between two surgeries varied from 5 to 6 months.

Only two cases were diagnosed as syndrome with craniofacial abnormalities. One child had Chromosome 9, partial monosomy, and other child had Muenke syndrome (FGFR3-related). In 23 of 24 cases (95.83%) an intraoperative or postoperative blood transfusion was required. Estimated blood loss was not routinely recorded due to immeasurable loss into and under surgical drapes.

In 24 patients treated there was no mortality. In one case with severe anterior plagiocephaly, postoperative subcutaneous hematoma in the affected side was developed, but this was successfully treated using only percutaneous puncture. For the same patient intraoperative dural tears occurred, but this was successfully treated during the same surgery.



**Fig. 3.** A. The patient with a severe form of scaphocephaly in the modified prone position. B. Craniolacunae and lines of osteotomy. C, D. After cranial vault remodeling, fixation with resorbable plates



**Fig. 4.** A. Osteotomy line in the case of scaphocephaly. B. Reshaped cranium, bone fragments fixed with titanium microplates

During early and late postoperative periods no infections, CSF leakage and dural tears were observed. No neurological impairments or any signs of neurological deficits were observed by any of treated patients.

In all of cases parents were satisfied with children's changed head shape and aesthetic results.

## REVIEW OF DIAGNOSTICAL METHODS AND DISCUSSION

In a typical case the morphological skull changes in each form of craniosynostosis are so specific that the diagnosis is essentially clinical and the difficulty of diagnosing craniosynostosis varies by case (10). In all clinical cases a careful medical history should be taken. Clinical and neurological examination is very important as craniosynostosis can result in raised intracranial pressure (11) and in untreated cases can lead to some neurological deficits or increased irritability (12). This can be seen especially in the cases of untreated scaphocephalies and trigonocephalies. Although in typical cases of craniosynostosis the diagnosis can be made after clinical evaluation, many surgeons tend to confirm diagnosis radiologically, especially in the cases of planned surgical treatment (13). Radiological evaluation is necessary to characterize the deformity, make the correct differential diagnosis and to guide the corrective surgical procedure (14).

Different instrumental methods in the diagnostics of craniosynostosis are or were used.

Historically, plain radiography has served as an initial imaging modality in a child with an abnormal head shape, and it remains a cost-effective method in infants with low risk of craniosynostosis (15). One of its benefits is that the plane radiography can be successfully done without general anesthesia opposite to the magnetic resonance imaging or computed tomography (16). The diagnosis of craniosynostosis is based on the primary (perisutural sclerosis, localized breaking, bony bridging, loss of visualization of the suture) and secondary signs (fingerprinting, beaten copper) visible on standard radiography (17). Also additional signs showing the type of compensatory growth or deformity can be seen in plain radiographs. Anteroposterior and lateral views of the skull are usual (18). It is important to evaluate the entire length of each suture because only a small segment may be in-

involved (19). Furthermore, primary radiographic findings are often unreliable in the first 3 months of life (20). Interpretation of plain radiographs is often challenging due to the low density of the skull in a neonate (21). In the majority of cases, however, evaluation of cranial sutures is inadequate with radiographs alone, so further investigations should be done (22).

Ultrasound is an effective, fast, low-cost, radiation-free method that requires no sedation in the management of children with head deformity (21). This examination is applicable only in cases with open fontanels, however, with a very high quality rating (7). Normal sutures show an uninterrupted hypoechoic gap between suture margins, while synostotic sutures show loss of this hypoechoic space and less specific signs include thickened margins, loss of beveled edges, and asymmetry of the fontanels (23). Several studies showed that ultrasound may be used in the prenatal diagnosis of synostosis (7, 24). Stelnicki et al. showed that standard ultrasonography of the calvarial sutures – in the absence of other craniofacial malformations – may be a feasible method for diagnosing simple, nonsyndromic craniosynostosis in utero but also with some diagnostic errors (7).

Magnetic resonance imaging (MRI) helps to detect cerebral and craniofacial soft-tissue anomalies seen in association with craniosynostosis, especially of syndromic variety (23). It is an excellent technique for the diagnosis of associated diseases of the cerebrum like, e. g. midline anomalies, lesions of the parenchyma, intracranial herniation and hydrocephalus (7). But in non-syndromic craniosynostosis, only MRI can be indicated in the cases of trigonocephaly because of its association with additional cerebral deformations (23). MRI is not a strong modality for evaluating bony abnormalities and thus cannot be used as the primary method of evaluating craniosynostosis (24).

Conventional Computed Tomography (CT) with three-dimensional (3D) reconstructions is considered the most complete and accurate imaging modality to diagnose craniosynostosis (26). The study of Vannier et al. demonstrated that three-dimensional shaded-surface reconstruction from CT scans is superior to conventional plain radiographs and CT scans in diagnosing craniosynostosis (27). CT with 3D reconstructions yields rich information on the alteration of the

normal skull shape and on the synostosed suture and it is most useful in planning the osteotomy and skull reshaping surgery (28). There are several indications for performing the computed axial tomographic scans. The first is to confirm a clinical impression of craniosynostosis by objectively demonstrating fusion of the suture. The second is to look for coexisting abnormalities, such as Arnold-Chiari malformation, which is frequently seen in Crouzon syndrome. The third is to visualize the extent of craniofacial bony disease (14). Some authors recommend to perform computed tomographic scanning for all patients with craniosynostosis to exclude hydrocephalus and other cranial or cerebral abnormalities (19).

However, with regard to the risk of radiation exposure particularly in young infants, CT scanning and even plain radiography should be indicated extremely carefully (29). Radiation exposure in children can result in greater cancer risk (30). Physicians, CT technologists, and health authorities should work together to minimize the radiation dose for children to as low as reasonably achievable and encourage responsible use of this essential diagnostic tool (31).

Premature fusion of the cranial sutures (craniosynostosis) may be associated with gross cranial or facial deformity, and may also cause serious constriction of the developing brain (craniostenosis), eyes (orbitostenosis) and facial viscera (faciostenosis) (32).

The main goal of the treatment of craniosynostosis is to restore normal appearance of the skull and increase the cranial volume so that the growing brain can be accommodated without any pressure effects on vital structures. The aim of surgical intervention is to excise the prematurely fused suture and correct the associated deformities of the calvaria, reshape the cranial vault and to perform fronto-orbital advancement to correct any recessed supraorbital rims (1). If the synostosis goes uncorrected, the deformity progresses to involve the facial skeleton, which is associated with asymmetry of the face and malocclusion. Therefore, the surgical goal is to increase the intracranial volume, especially under the fused suture, and prevent any long-term complications (5).

The first surgery on craniosynostoses was described by Marie-Lannelongue in 1890 (33). At that time mostly simple craniectomy was per-

formed. Simple craniectomy was unfortunately accompanied by a high rate of reossification and gave only modest results, unless mobilization of the orbits, midface, and cranium was performed concurrently (34). Today there are many surgical techniques and modifications that have been described for skull reconstruction in craniosynostosis. Two main groups of surgery for craniosynostosis can be distinguished. The first one is calvarial vault remodeling and the second is minimally invasive endoscopic assisted surgery. During the vault remodeling the skull bones can be divided into many parts and these parts may be rearranged to give a natural appearing shape. Some areas may need to have onlay bone grafting by using split cranial bone grafts (1). Performing endoscopic assisted surgery only the affected suture and the adjacent bone area are removed and the calvarian form is corrected later using a cranial helmet. This technique can be used not in all cases. Early diagnosis is very important for planning a surgical repair of craniosynostosis (35).

The optimal timing for the surgical treatment of craniosynostosis is still controversial.

Many craniosynostosis series have been reported in literature. Based on Utria et al., the ideal operative for craniosynostosis appears to be 6–9 months of age when there is no concern for elevated intracranial pressures (35).

A delay in surgery beyond the first 9 to 12 months of life leads to progressive deformity of the cranial base, resulting in abnormal facial growth and asymmetry of the maxilla and mandible (35). The best time to intervene is when the infant is between 3 and 9 months of age (37) to take advantage of this period of rapid brain and skull growth, to provide an optimal chance for reossification of the surgical cranial defects, and to ensure ease of bone remodeling (13). The calvaria in a child 3 to 9 months of age is still malleable and, therefore, quite easy to shape, (37) but one large-scale study reported that younger age (<9 months) can be considered as a predictor factor for surgery complications (38). General indications for surgical intervention in nonsyndromic craniosynostosis include the presence of cosmetic deformity and/or functional impairment, such as intracranial hypertension or optic atrophy (39).

In our cases the mean age at the time of surgery was  $13.47 \pm 8.2$  months, but eliminating 3 patients

whose age at the time of surgery was over 2 years, the mean age of other 21 patients was  $10.63 \pm 1.77$  months.

The relatively big mean age at the time of surgery was caused by late referral of these patients to the craniofacial surgeons. The main factors, in our opinion, were and still are poor or insufficient physician's knowledge about craniosynostosis and the anxiety or even fear of any bigger/modern craniosynostotic or craniofacial surgery. After we started our activity as a craniofacial team and gave lectures for pediatricians and other physicians, also showed the results of our treatment, the amount of timely referred and as well as treated patients started to grow up.

Other authors also noted that bigger amount of surgeries was caused by education of pediatricians, neurologists and other physicians who can have any contacts with these patients up to 12 months. During this time, cerebral growth is greatest and, in craniosynostosis, effects on the brain may be detrimental (33).

Most often complications after open surgical repair of craniosynostosis include a moderate amount of blood loss during the surgery, infection (3–6%) (40), cerebrospinal fluid leak, meningitis. The mortality has been reported to be around 0.1–2.2% (41). This depends on patient selection and patient series. Patients with non-syndromic craniosynostosis, if operated early, have a very good postoperative outcome and the need for the second surgery is minimal (1). However, in syndromic cases the reoperation rates are quite high, to the tune of about 2–13% (41). It was stated that the highest number of complications was related to complete cranial vault remodeling (holocranial dismantling) in scaphocephalies and multiple synostosis and after the use of internal osteogenic distractors (43).

Harrop et al. reported a morbidity of 0.02% and no mortality in 40 consecutive craniosynostosis operated cases. Kadri and Mawla referred a mortality of 3 patients (2.58%) in a study of 116 children with cranio-synostosis. Nonaka et al., in a series of 25 patients with craniosynostosis, had a morbidity of 12% (3 patients). Ferreira et al. presents the morbidity 9.7% and mortality 2.6% (34). In the last cases of this series, it was observed that surgical time, blood loss in the trans-operative and post-operative periods, postoperative com-

plications and mortality rates showed a significant reduction (34).

Our study also showed that the last surgeries were performed in shorter time than the first surgeries although it was statistically insignificant ( $p > 0.05$ ).

These findings may reflect an improvement in the relationship among surgical (craniomaxillofacial, neurosurgical and anesthesiological) team members in dealing with this complex and interesting disease (34).

According to the literature the most frequent complication was non-filiated postoperative hyperthermia (13.17% of the cases) followed by infection (8.10%), subcutaneous haematoma (6.08%), dural tears (5.06%) and cerebrospinal fluid (CSF) leakage (2.7%) (43).

In our last treated cases we have used Manitol (0.5–1 g/kg) to reduce intracranial hypertension and cerebral edema during the division of the frontoorbital bandeau. In our opinion, this maneuver helps to reduce possible brain damaging caused by direct pressure exposing the anterior cranial fossa. However, more experience is required to confirm this statement as well the usage of tranexamic acid to reduce intraoperative blood loss.

A multidisciplinary approach, including neurosurgeons, neurologists, and pediatricians, and appropriate training of the clinical surgical staff can minimize the risks and decrease the complications in the treatment of craniosynostosis, leading to a satisfactory outcome (34).

In our case series we had a case of subcutaneous haematoma and dural tears (4.17%). We had no postoperative infection and any mortality but it is difficult to compare due to a relatively small amount of treated cases.

It should be noted that the craniofacial team is composed not only of a cranio-maxillofacial surgeon and a neurosurgeon. An anesthesiologist also plays a very important role during the intra- and postoperative treatment of craniosynostotic patients. Specific anesthetic challenges occur during this type of operation. The main problems for an anesthesia team are that craniofacial surgery may be associated with sudden cardiovascular changes and with the potential for significant blood loss (44). High-risk groups for bleeding include those with weight  $<10$  kg, age  $<18$  months, craniofacial syndromes, pansynostosis, operating time more



than five hours and patients with known high intracranial pressure. In a modified prone position (sphinx), hyperextension of the neck may result in spinal cord and orbital injury. The anesthesiologist should attempt to minimize factors that increase intracranial pressure, such as hypercapnia and hypoxia, and factors that increase venous pressure, such as the patient's position and coughing.

Craniosynostosis should be ideally managed in a multidisciplinary team. It is stated that objective evaluation of the results in craniofacial surgery constitutes a difficult issue and the end-results are still a subjective measure (45). Despite that, regular follow-up visits after surgery to monitor head growth, check for possible persistent craneolacuniae or increased intracranial pressure are necessary. Only regular follow-up can ensure that possible re-fusing of sutures will be diagnosed early and possible reoperation can be performed at the right time.

## CONCLUSIONS

Cranioplasties for correction of craniosynostosis give good aesthetic results and this is quite a safe method which helps to correct the head shape as well as improves the social adaptation of patients. Final evaluation of the performed cranioplasties requires a longer observation period.

Received 13 April 2015

Accepted 26 May 2015

## References

- Sharma RK. Craniosynostosis. *Indian J Plast Surg.* 2013; 46(1): 18–27.
- Lima D. The management of deformational plagiocephaly: a review of the literature. *J Prosth Orthot.* 2004; 16(4): S9–14.
- Stephen MW. The pathogenesis of craniosynostosis in the fetus. *Yonsei Med J.* 2001; 42: 646–59.
- Speltz ML, Kapp-Simon KA, Cunningham M, Marsh J, Dawson G. Single suture craniosynostosis: a review of neurobehavioral research and theory. *J Pediatr Psychol.* 2004; 29(8): 651–68.
- Panchal J, Uttchin V. Management of craniosynostosis. *Plast Reconstr Surg.* 2003; 111(6): 2032–48.
- Guyuron B. *Plastic Surgery: Indications and Practice.* Saunders; 2009.
- Kotrikova B, Krempien R, Freier K, Muhling J. Diagnostic imaging in the management of craniosynostoses. *Eur Radiol.* 2007; 17: 1968–78.
- Zinkuviene A. Respublika. Gyvenimas. 2009 March 9. Lithuanian.
- Van der Meulen JJ, Nazir PR, Mathijssen IM, van Adrichem LN, Ongkosuwito E, Stolk-Liefferink SA, Vaandrager MJ. Bitemporal depressions after cranioplasty for trigonocephaly: a long-term evaluation of (supra) orbital growth in 92 patients. *J Craniofac Surg.* 2008; 19(1): 72–9.
- Vannier MW, Pilgram TK, Marsh JL, Kraemer BB, Rayne SC, Gado MH. Craniosynostosis: diagnostic imaging with three-dimensional CT presentation. *AJNR Am J Neuroradiol.* 1994; 15: 1861–9.
- Massimi L, Caldarelli M, Tamburrini G, Paternoster G, Di Rocco C. Isolated sagittal craniosynostosis: definition, classification, and surgical indications. *Childs Nerv Syst.* 2012; 28(9): 1311–7.
- Boltshauser E, Ludwig S, Dietrich F, Landolt MA. Sagittal craniosynostosis: cognitive development, behaviour, and quality of life in unoperated children. *Neuropediatrics.* 2003 Dec; 34(6): 293–300.
- Sze RW, Hopper RA, Ghioni V, Gruss JS, Ellenbogen RG, King D, et al. MDCT Diagnosis of the Child with Posterior Plagiocephaly. *AJR Am J Roentgenol.* 2005; 185: 1342–6.
- Persing JA. Treatment of Craniosynostosis. *Plast Reconstr Surg.* 2008; 121(4): 1–11.
- Medina LS, Richardson RR, Crone K. Children with suspected craniosynostosis: a cost-effectiveness analysis of diagnostic strategies. *AJR Am J Roentgenol.* 2002; 179: 215–21.
- Anderson PJ, Yong R, Surman TL, Rajion ZA, Ranjitkar S. Application of three-dimensional computed tomography in craniofacial clinical practice and research. *Aust Dent J.* 2014; 59(1): 174–85.
- Benson ML, Oliverio PJ, Yue NC, Zinreich SJ. Primary craniosynostosis: imaging features. *AJR Am J Roentgenol.* 1996; 166: 697–703.
- Kabbani H, Raughveer ST. Craniosynostosis. *Am Fam Physician.* 2004; 69: 2863–70.
- Aviv R, Rodger E, Hall C. Craniosynostosis. *Clin Radiol.* 2002; 57: 93–102.
- Fernbach SK, Naidich TP. Radiological evaluation of craniosynostosis. In: Cohen MM Jr, editor. *Craniosynostosis: diagnosis, evaluation, and management.* New York: Raven Press; 1986.
- Simanovsky N, Hiller N, Koplewitz B, Rozovsky K. Effectiveness of ultrasonographic evaluation of the

- cranial sutures in children with suspected craniosynostosis. *Eur Radiol.* 2009; 19: 687–92.
22. Khanna PC, Thapa MM, Iyer RS, Prasad SS. Pictorial essay: The many faces of craniosynostosis. *Indian J Radiol Imaging.* 2011; 21(1): 49–56.
23. Badve CA, Mallikarjunappa MK, Iyer RS, Ishak GE, Khanna PC. Craniosynostosis: imaging review and primer on computed tomography. *Pediatr Radiol.* 2013; 43: 728–42.
24. Khan MA, Weltman DI, Glasier CM, Coombs BD, Hernanz-Schulman M, Krasny RM, et al. Craniosynostosis imaging [Internet]. 2013 Jun 7. Available from: <http://emedicine.medscape.com/article/407856-overview>
25. Lumenta CB, Haase J, Rocco CDi, Mooij JJA. *Neurosurgery.* Springer; 2010.
26. Tartaro A, Larici AR, Antonucci D, Merlino B, Colosimo C, Bonomo L. Optimization and diagnostic accuracy of computerized tomography with tridimensional spiral technique in the study of craniosynostosis. *Radiol Med.* 1998; 96(1–2): 10–7.
27. Vannier MW, Hildebolt CF, Marsh JL, Pilgram TK, McAlister WH, Shackelford GD, et al. Craniosynostosis: diagnostic value of three-dimensional CT reconstruction. *Radiology.* 1989; 173: 669–73.
28. Ciurea AV, Toader C, Mihalache C. Actual concepts in scaphocephaly. *J Med Life.* 2011; 4(4): 424–31.
29. Schweitzer T, Böhm H, Meyer-Marcotty P, Collmann H, Ernestus RI, Krauß J. Avoiding CT scans in children with single-suture craniosynostosis. *Childs Nerv Syst.* 2012; 28(7): 1077–82.
30. Brenner D, Elliston C, Hall E, Berdon WE. Estimated risks of radiation-induced fatal cancer from pediatric CT. *AJR Am J Roentgenol.* 2001; 176: 289–96.
31. Chodick G1, Ronckers CM, Shalev V, Ron E. Excess lifetime cancer mortality risk attributable to radiation exposure from computed tomography examinations in children. *Isr Med Assoc J.* 2007; 9(8): 584–7.
32. David DJ. Advances in the management of the craniosynostoses. *ANZ J Surg.* 2003; 73: 949–57.
33. 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(4): 103–13.
34. Ferreira MP, Collares MV, Ferreira NP, Kraemer JL, Pereira Filho Ade A, Pereira Filho Gde A. Early surgical treatment of nonsyndromic craniosynostosis. *Surg Neurol.* 2006; 65 Suppl 1: S22–6.
35. Utria AF, Mundinger GS, Bellamy JL, Zhou J, Ghasemzadeh A, Yang R, et al. The importance of timing in optimizing cranial vault remodeling in syndromic craniosynostosis. *Plast Reconstr Surg.* 2014.
36. Marsh JL, Vannier MW. Cranial base changes following surgical treatment of craniosynostosis. *Cleft Palate J.* 1986; 23 Suppl 1: 9.
37. Panchal J, Uttchin V. Management of craniosynostoses. *Plast Reconstr Surg.* 2003; 111: 2032–48.
38. Lee HQ, Hutson JM, Wray AC, Lo PA, Chong DK, Holmes AD, et al. Analysis of morbidity and mortality in surgical management of craniosynostosis. *J Craniofac Surg.* 2012; 23: 1256–61.
39. Renier D, Lajeunie E, Arnaud E, Marchac D. Management of craniosynostoses. *Childs Nerv Syst.* 2000; 16(10–11): 645–58.
40. Fearon JA, Yu J, Bartlett SP, Munro IR, Chir B, Whitaker L. Infections in craniofacial surgery. A combined report of 567 procedures from two centers. *Plast Reconstr Surg.* 1997; 100: 862–8.
41. Czerwinski M, Hopper R, Gruss J, Fearon J. Major morbidity and mortality rates in craniofacial surgery: An analysis of 8,101 major procedures. *Plast Reconstr Surg.* 2010; 126: 181–6.
42. McCarthy J, Glasberg S, Cutting C, Epstein F, Grayson B, Ruff G, et al. Twenty-year experience with early surgery for craniosynostosis: I. Isolated craniofacial synostosis: Results and unsolved problems. *Plast Reconstr Surg.* 1995; 96: 272–83.
43. Esparza J, Hinojosa J. Complications in the surgical treatment of craniosynostosis and craniofacial syndromes: apropos of 306 transcranial procedures. *Childs Nerv Syst.* 2008; 24: 1421–30.
44. White N, Bayliss S, Moore D. Systematic review of interventions for minimizing perioperative blood transfusion for surgery for craniosynostosis. *J Craniofac Surg.* 2015; 26(1): 26–36.
45. Esparza J, Hinojosa J, García-Recuero I, Romance A, Pascual B, Martínez de Aragón A. Surgical treatment of isolated and syndromic craniosynostosis. Results and complications in 283 consecutive cases. *Neurocirugía.* 2008; 19: 509–29.

Linus Zaleckas, Arijanda Neverauskienė,  
Vidūnas Daugelavičius,  
Dominika Šidlovskaitė-Baltakė, Ramūnas Raugalas,  
Berta Vištartaitė, Evelina Balčiūnaitė

## KRANIOSINOSTOZIŲ DIAGNOSTIKA IR GYDYMAS: VILNIAUS KOMANDOS PATIRTIS

### *Santrauka*

**Tikslas.** Apžvelgti kraniosinostozinių diagnostikos metodus, peržiūrėti Vilniaus (Lietuvos) komandos kraniosinostozinių chirurginio gydymo bei anestezijos patirtį, metodus ir pateikti ankstyvuosius chirurginio gydymo rezultatus.

**Medžiaga ir metodai.** Retrospektyviniame tyrime įvertinti visi pacientai, kurie dėl įvairaus tipo kraniosinostozinių buvo operuoti nuo 2009 m. sausio 1 d. iki 2014 m. gruodžio 31 d. Vertintas pacientų amžius operacijos metu, deformacijos tipas, chirurginė technika, anestezijos metodai ir eiga, intra- ir pooperacinės komplikacijos, galvos forma, tėvų pasitenkinimas.

**Rezultatai.** Iš viso operuoti 24 pacientai. Pacientų amžiaus vidurkis operacijos metu buvo  $13,47 \pm 8,2$  mėnesiai (min 7,3; maks 46,5), tačiau atmetus 3 pacientus, kuriems operuojant buvo per 2 metus (atitinkamai 24,5; 29 ir 46 mėnesiai), likusių (21 paciento) amžiaus vidurkis siekė  $10,63 \pm 1,77$  (min 7,3; maks 14,1) mėnesius. Visiems 24 pacientams diagnozė buvo nustatyta kliniškai

ir vėliau patvirtinta kompiuterinės tomografijos tyrimu. 9 pacientai (37,5 %) operuoti dėl izoliuotos trigonicefalijos, 7 (29,2 %) – dėl izoliuotos skafocfalijos, 7 (29,2 %) – dėl izoliuotos priekinės plagiocefalijos ir 1 pacientas – dėl kombinuotos užpakalinės plagiocefalijos ir skafocfalijos. 2 pacientams diagnozuotos sindrominės kraniosinostozės. Visais atvejais atlikta stabili (rigidiška) osteosintezė. Operacijų trukmės vidurkis –  $336,47 \pm 59,63$  minutės (min 308,13; maks 364,82). Po operacijos pacientai intensyviosios terapijos skyriuje vidutiniškai praleido  $2,53 \pm 1,28$  dienas (min 1,92; maks 3,14). 23 pacientams (95,83 %) buvo atlikta intraoperacinė arba pooperacinė kraujo transfuzija. Ankstyvuojanti ir vėlyvuojanti pooperacinė infekcija nebuvo užfiksuota, cerebrospinalinio skysčio nutekėjimo ar smegenų dangalo pakenkimo atvejų. Nė vienam tirtam pacientui po operacijos nepasireiškė neurologiniai sutrikimai ar neurologinis deficitas, nebuvo nė vieno mirties atvejo. Visais tirtaisiais atvejais pacientų tėvai buvo patenkinti pasikeitusia galvos forma ir chirurginio gydymo estetiniu rezultatu.

**Išvados.** Kranioplastika yra saugus kraniosinostozinių gydymo metodas, leidžiantis pasiekti gerą estetinių gydymo rezultatų, koreguojantis galvos formą bei palengvinantis pacientų socialinę adaptaciją.

**Raktažodžiai:** kraniosinostozė, kompiuterinė tomografija, kraniofacialinė deformacija, kraniofacialinė chirurgija, vaizdavimas