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# Surgical correction of lambdoid synostosis – New technique and first results $^{\star}$



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#### ABSTRACT

*Objective:* Premature craniosynostosis of the lambdoid suture is rare. The use of differential diagnosis to rule out positional occipital plagiocephaly is crucial. Nevertheless, once diagnosed, lambdoid craniosynostosis requires corrective surgery to prevent intracranial harm and aesthetic stigma by significant dyscrania. Operative correction of the lambdoid fusion is often performed by suturectomy and helmet therapy, total occipital remodeling interventions, transposition of occipital bone flaps, or occipital advancement procedures either with or without distraction osteogenesis. We present a simple surgical maneuver to potentially correct the occipital and suboccipital constriction caused by unilateral lambdoid craniosynostosis.

*Materials and methods:* Three patients with true unilateral lambdoid synostosis underwent surgery. A straight-line skin incision was created, beginning at the caudal pole of the ipsilateral mastoideal bulge. The incision ran cranially and corresponded to the course of the lambdoid suture up to the posterior fontanel. The periosteum was incised and the contralateral (potent) lambdoid suture was identified at its origin. One burr-hole was created to separate the dura from the intern tabula. Afterwards, a square meander-shaped craniotomy was performed along the assumed course of the lambdoid suture. The squares were then forced apart to form the occipital and suboccipital area into a more rounded shape. The squares were fixed in those positions with resorbable plates or sutures. Pre- and postoperative three-dimensional (3D) photoscans were performed and analyzed with special software to follow the perioperative course of the cranial shape.

*Results:* This new approach minimized the operative time and degree of blood-loss, and rounding of the occipital area was accomplished with only one unilateral intervention. In terms of the available photogrammetric data of two of the patients, the cranial vault asymmetry index, posterior symmetry ratio, and posterior/anterior skull volume ratio were improved but not normalized completely. The measurement outcome, as determined by an automated analysis of the photoscans, however, indicated clear flaws with regard to repeatability.

*Conclusion:* A unilateral approach using a square meander-shaped craniotomy and subsequent inclination may be a suitable surgical method for correcting assorted cases of lambdoid craniosynostosis. Transposition of the occipital bone flaps, subtotal craniectomies of the occipital area, and occipital advancements with or without distraction devices may not be essential in all cases of lambdoid synostosis. However, the reliability of the automated analysis of three-dimensional photoscans must be determined.

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- Abbreviations: CVAI, cranial vault asymmetry index; CI, cranial index; CVA, cranial vault asymmetry; OR, operation room; PAR, ratio of posterior/anterior skull volume (Q3 + Q4)/(Q1 + Q2); PSR, posterior symmetry ratio (Q3/Q4); ULS, unilateral lambdoid synostosis.
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#### 1. Introduction

Lambdoid craniosynostosis is a very rare condition with a prevalence of approximately one in every 40,000 births (David and Menard, 2000). Misdiagnosis is frequent (Cohen and MacLean, 2000) and is due mainly to the high frequency of positional occipital flattening following the "back-to-sleep-campaign"

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begun in 1992 (AAP, 1992; Mawji et al., 2013; Turk et al., 1996). If true lambdoid synostosis is encountered, surgery is the only option for correcting intra- and extracranial constriction. Surgical methodology and timing varies. A recently published review of the surgical correction of unilateral lambdoid synostosis (ULS) systematically evaluated the available published data and found that the number of presented cases in the literature with true ULS was as low as 188 and that the surgical modus operandi was heterogeneous (Al-Jabri and Eccles, 2014). Additionally, the preoperative and follow-up diagnostic criteria regarding ULS were critically reviewed by the authors. David et al. defined four characteristic clinical patterns for ULS (David and Menard, 2000): mastoideal bulging, an ipsilateral inferior tilt of the skull base, a trapezoid head shape from the vertex view, and contralateral posterior parietal bossing. Furthermore, the occiput is shaped like a parallelogram when observed from behind (Fig. 1a, b) and the parietooccipital region shows a retraction perpendicular to the lambdoid suture running from the skull base over the inion towards the squamous suture.

Surgical correction is not always able to adjust to every instance of asymmetry (Komuro et al., 2004; Smartt et al., 2007). This is particularly the case with regard to the cranio-caudal shift of the cranial base and the consecutive inferior positioning of the ipsilateral external auricular meatus and external ear. Jimenez and Barone et al. advocate endoscopic strip suturectomy and helmet therapy as the method of choice for lambdoid synostosis and report a short period of hospitalization and minimal blood loss (Jimenez et al., 2002; Johnson et al., 2000). Most other authors, however, tend to prefer open coronal approaches with bone-flap-remodeling or switching (Elliott et al., 2013; Liu et al., 2008; Persing et al., 1988; Sgouros et al., 1996), barrel-staving osteotomies (Jimenez and Barone, 1995), distraction osteogenesis (Komuro et al., 2004), or spring-assisted correction of the cranial deformity (Arnaud et al., 2012), among others.

In this study, we present an endorsement of the common techniques used to correct ULS and provide objective anthropometric results collected pre- and postoperatively by examiner-independent photogrammetry (Wilbrand et al., 2012).

#### 2. Materials and methods

Children with ULS normally undergo surgery between the sixth and eighth month of life. We performed preoperative threedimensional (3D) computed tomography (CT) scanning to provide diagnostic certainty. Additionally, a 3D photogrammetric scan of the head was performed one day before the operation and approximately four weeks after.

#### 2.1. Operative technique

After removal of the hair, patients were placed in a 45° position contralateral to the synostosis. Following the additional application of a local anesthetic and the creation of a sterile environment, a straight-line skin incision was performed that ran from the onset of the nuchal musculature of the affected side upwards to the assumed position of the posterior fontanel (Fig. 2). An epiperiosteal dissection was made anteriorly and posteriorly and the origin of the contralateral and potent lambdoid suture was then identified (Fig. 3). The periosteum was incised downwards along the assumed course of the ipsilateral and synostosized lambdoid suture, from the origin of the contralateral lambdoid suture into the ipsilateral suboccipital region. Thereafter, a square meander-shaped craniotomy was performed over the entire course of the synostosized suture after releasing the dura from the tabula interna over a cranially and eventually caudally obtained burr-hole. Decompressing straight-line osteotomies were additionally obtained perpendicular to the vertex of the meander (Fig. 4). The craniotomized bone flaps were then forced apart, microfractured, and remodeled with Tessier forceps. Once a satisfactory rounding of the restricted area was obtained, the divided bone flaps were fixed in the distracted position with slowly resorbing suture osteosynthesis (Fig. 5) (Fearon, 2003). Finally, a drain was inserted and the skin wound closed in multilavers.

Children undergoing this procedure received blood transfusions as a precaution, rather than to counteract complications arising from severe bleeding. Postoperatively, patients remained in the intensive care unit (ICU) overnight and were transferred to the



Fig. 1. (a): Clinical picture of ULS, parallelogram-shaped head deformity from posterior view in Child 2. (b): Typical head shape in lambdoid synostosis.

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