Management of Lambdoid Craniosynostosis: A Comprehensive and Systematic Review

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Keywords

Lambdoid craniosynostosis · Neurosurgery · Craniofacial · Pediatrics · Plastic surgery

Abstract

Background: Craniosynostosis is a condition characterized by the premature fusion of 2 or more skull bones. Craniosynostosis of the lambdoid suture is one of the rarest forms, accounting for 1–4% of all craniosynostoses. Documented cases are separated into simple (single suture), complex (bilateral), and associated with adjacent synostoses (“Mercedes Benz” Pattern) or syndromes (i.e., Crouzon, Sathre-Chotzen, Antley-Bixler). This condition can manifest phenotypic deformities and neurological sequelae that can lead to impaired cognitive function if improperly treated or left undiagnosed. Preferred surgical techniques have varied over time but all maintain the common goals of establishing proper head shape and preventing of complications that could contribute to aforementioned sequelae. **Summary:** This comprehensive review highlights demographic distributions, embryological development, pathogenesis, clinical presentation, neurological sequelae, radiologic findings, surgical techniques, surgical outcomes, and postoperative

considerations of patients with lambdoid craniosynostosis presentation. In addition, a systematic review was conducted to explore the operative management of lambdoid craniosynostosis using PubMed, Embase, and Scopus databases, with 38 articles included after screening. **Key Messages:** Due to a low volume of published cases, diagnosis and treatment can vary. Large overlap in presentation can occur in patients that display lambdoid craniosynostosis and posterior plagiocephaly, furthering the need for comprehensive analysis. Possessing the knowledge and tools to properly assess patients with lambdoid craniosynostosis will allow for more precise care and improved outcomes.

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Introduction

The earliest documented case of isolated lambdoid craniosynostosis was a left lambdoid synostosis in an immature human fossil dated at a minimal age of 530,000 years [1]. Currently, the prevalence of true lambdoid craniosynostosis remains low, contributing to 1–4% of all craniosynostoses [2]. The exact etiology of the condition is not known; however, a genetic predisposition is suggestive, as there have been documented cases of familial [3]

Table 1. Delineated search terms used in the systematic review

Database	Search terms
PubMed	{("cranosynostoses"[MeSH] AND "lambdoid*") OR "lambdoid cranosynostosis*" OR "lambdoid synostosis*" OR "lambdoidal cranosynostosis*" OR "lambdoidal synostosis*" OR ("lambdoid*" AND ["cranosynostosis*" OR "synostosis*"])} An asterisk (*) represents truncation of the search term.
Scopus	{"Lambdoid cranosynostosis*" OR "lambdoid synostosis*" OR "lambdoidal cranosynostosis*" OR "lambdoidal synostosis*" OR ("lambdoid*" AND ["cranosynostosis*" OR "synostosis*"])}:ti,ab,kw
Embase	TITLE-ABS-KEY{("lambdoid cranosynostosis*" OR "lambdoid synostosis*" OR "lambdoidal cranosynostosis*" OR "lambdoidal synostosis*" OR ["lambdoid*" AND ["cranosynostosis*" OR "synostosis*"]])}

and dizygotic twins [4] lambdoid cranosynostoses. A clinical presentation of true lambdoid cranosynostosis is highly variable given a large degree of overlap with deformational posterior plagiocephaly, complicating diagnosis. Common physical examination findings include frontal bossing, posterior plagiocephaly, and changes in ear position, torticollis, and trapezoidal head shape [5]. Because of the variability in presentation, confirmation of diagnosis is obtained with computed tomography (CT) scan and can guide the surgeon toward the proper surgical intervention. Initially, open-strip craniectomy was the common treatment method; however, complications from the procedure and development of new techniques have promoted open cranial vault reconstruction and endoscopic suturectomy to the gold standard for surgical management. Postoperative considerations for patients with lambdoid cranosynostosis include monitoring of intracranial pressure (ICP), lab value maintenance, and implementation of helmet-molding orthoses to prevent suture refusion. A thorough evaluation and attention to detail at every step is necessary for a favorable outcome in a patient with lambdoid cranosynostosis.

Methods

A systematic review regarding the management of isolated lambdoid cranosynostosis was performed in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analysis (PRISMA) 2020 guidelines [6]. No protocol was registered. PubMed MEDLINE (National Library of Medicine), Embase (Elsevier), and Scopus (Elsevier) were searched on June 18, 2021, using keywords including lambdoid cranosynostosis with no restrictions on language, date, or article type (Table 1). Figure 1 demonstrates a PRISMA flowchart summarizing article inclusion (Fig. 1).

After the search was completed, duplicates were removed utilizing the automated deduplication function in Endnote (Clarivate

Analytics, London, UK). All remaining articles were screened by title and abstract for relevance. Articles progressing to full-text review were screened based on prespecified inclusion and exclusion criteria. Inclusion criteria were published in or translated into the English language, with full-text available, providing primary data, population of patients with isolated unilateral or bilateral lambdoid cranosynostosis, and discussing management and associated patient outcomes. Exclusion criteria were conference abstracts, commentaries, letters to the editor, reviews, meta-analyses, other subtypes of cranosynostosis, and lack of outcomes of management. Studies discussing multiple subtypes of cranosynostosis were included if outcomes specific to isolated lambdoid cranosynostosis patients were provided. A second reviewer replicated the search strategy and screening. Disagreements were reconciled by consensus.

After the set of included articles was finalized, studies were reviewed for characteristics designated a priori such as bibliographic data, aim, design, study participants, operative and postoperative details, outcomes, and length of follow-up. Outcomes of interest were blood loss, blood transfusion, major complications, reoperation, and head contour. The World Bank income classification was used to designate the income status countries of origin for all included studies [7]. The quality of each study was evaluating using the GRADE framework [8]. The risk of bias for each included study was determined using the ROBINS-I tool [9]. A judgment on the overall risk of bias was leveled by considering the risk of bias of all included studies in aggregate.

Results

A search of the literature resulted in 38 articles published from 1984 to 2021 that met eligibility criteria [5, 10–46]. Out of the 38 total articles, 32 (84.2%) were case series, 5 (13.2%) were retrospective cohort analyses, and 1 (2.6%) was a case report. In total, 433 patients were reported to have received surgical intervention for either unilateral or bilateral lambdoid cranosynostosis, with 1 article not reporting these data [37]. In the studies that included separate data for both isolated and bilateral syn-

Fig. 1. PRISMA flowchart outlining inclusion and exclusion criteria for systematic review.

ostoses, 268 (88.4%) isolated and 35 (11.6%) bilateral lambdoid craniosynostoses were recorded.

The most common surgical intervention utilized was strip craniectomy, with other cited articles utilizing “Sand Dollar and Staves” technique, posterior vault remodeling, sunrise technique, and distraction osteogenesis. The mean blood loss ranged from 7.5 to 640.5 mL and mean postoperative blood transfusions ranged from 0 to 117 mL. Head contour results showed improvement in entire cohort for 18 (69.2%) articles with 8 (31.8%) mentioning no change in parameters such as cranial fossa size, residual occipital deformities, hemifacial deficiencies, posterior fossa deflection, petrous ridge asymmetry, and anteroinferior ipsilateral ear displacement. The rate of complication ranged from 0% to 20% but never exceeded 1 patient per cohort. The rate of reoperation ranged from 0% to 23.8%, with a maximum of 5 patients requiring reoperation in 1 cohort. The average length of hospitalization ranged between 1 and <5 days. The mean length of follow-up ranged from 7 months to 84 months with 50% (19/38) of articles documenting these data. The quality of

evidence was low in most studies. Given that most included studies had a high risk of bias, the risk of bias in this systematic review was high overall (Table 2).

Discussion

Etiology and Pathogenesis

Craniosynostosis as a disease entity is fairly common, affecting 1 in 2,100–2,500 births [47–49]. Approximately 85% of children with craniosynostosis are asymptomatic (e.g., have isolated craniosynostosis), 7% are symptomatic, and 9% show symptoms of syndromic disease [50, 51]. Lambdoid craniosynostosis is the rarest variant, accounting for approximately 2% of all craniosynostosis cases and affecting 1 in 33,000 infants [5, 52–54]. According to a recent large case series, there appears to be no predilection for gender (52% male, 48% female) or laterality (52% left sided, 48% right sided).

In the developing embryo, the cranial vault forms from the mesenchymal tissue that is initially arranged as a cap-

Table 2. Studies included in the systematic review

Paper	Country	Income status	Study design	Risk of bias	Quality grade	Isolated lambdoid craniosynostosis undergoing surgery (unilateral/bilateral), n	Mean age at diagnosis, months	Mean age at surgery, months	Type of surgery	Mean operative time, h	Mean blood loss, mL	Mean postoperative transfusion, units, mL	Hospitalization length, days	Major complications, N (%)	Reoperation, N (%)	Head contour	Mean length of follow-up, months	Key findings
Abu-Ghname et al. [10]	USA	HIC	Case series	High	Low	4 (4/0)	9	12.3	Sand Dollar and Staves	2.5	70	0	2.67	0 (0)	NR	NR	10.6	Decreased operative time, blood loss, and hospital stay with satisfactory aesthetic outcomes with this approach
Barone and Jimenez [11]	USA	HIC	Case series	High	Low	2 (2/0)	NR	5	Endoscopic strip craniectomy	NR	7.5	0	<2	NR	NR	Cranial remodeling	NR	Endoscopic strip craniectomy allows skull remodeling with less blood loss, less blood transfused, less operative time, and last expense
Baumgartner et al. [12]	USA	HIC	Case series	High	Low	1	NR	2	Microscope	0.92	20	NR	3	NR	NR	Posterior asymmetry went from 5 mm preoperatively to 2 mm postoperatively to 0 after helmet therapy	NR	Microscopic approach is best suited for infants with lambdoid craniosynostosis
Borad et al. [5]	USA	HIC	Case series	High	Low	25	9.6	10.4	Posterior calvarial remodeling with/without barrel stave osteotomies and full thickness calvarial bone grafts	NR	NR	NR	NR	0 (0)	8	All improved	43.8	Surgery leads to safe and consistent long-term results Patients with concomitant torticollis may have residual facial asymmetry and head shape abnormalities despite optimal surgical correction of the lambdoid synostosis
Cartwright et al. [13]	USA	HIC	Case series	High	Low	6	4.3	NR	Endoscopic strip craniectomy	1.37	33.6	0	1	0 (0)	NR	NR	NR	Most patients were normocephalic or nearly postoperatively
Collett et al. [14]	USA	HIC	Retrospective cohort	Moderate	Moderate	12	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	Limited evidence of broad deficits in executive function or attention in children with single suture craniosynostosis relative to controls Neurodevelopment in children with single suture craniosynostosis is variable
Dias et al. [15]	USA	HIC	Retrospective cohort	Moderate	Moderate	30 (30/0)	3.2	7.3	Strip craniectomy	NR	NR	NR	NR	NR	NR	Anterior and posterior asymmetries improved but persisted relative to controls	21.6	Improvements following unilateral lambdoid craniectomy are difficult to interpret in isolation

Table 2 (continued)

Paper	Country	Income status	Study design	Risk of bias	Quality grade	Isolated lambdoid craniostosis undergoing surgery (unilateral/bilateral), n	Mean age at diagnosis, months	Mean age at surgery, months	Type of surgery	Mean operative time, h	Mean blood loss, mL	Mean postoperative transfusion, units, mL	Hospitalization length, days	Major complications, N (%)	Reoperation, N (%)	Head contour	Mean length of follow-up, months	Key findings
Elliott et al. [16]	USA	HIC	Case series	High	Low	5 (5/0)	NR	16	Posterior vault remodeling	NR	NR	NR	NR	NR	NR	Cranial fossa area remained enlarged relative to the ipsilateral side, contralateral middle fossa became more retrodisplaced, ear position also remained asymmetrical	21.8	Cranioplasty effectively restores calvarial shape but does not significantly alter the dysmorphic features in endocranium
Fearon et al. [17]	USA	HIC	Case series	High	Low	25 (25/0)	NR	12	Posterior vault remodeling	NR	NR	NR	2.3	0 (0)	2 (10)	NR	68.4	60% developed chiari malformations
Fearon et al. [18]	USA	HIC	Case series	High	Low	1 (1/0)	NR	NR	Open remodeling	NR	NR	NR	NR	NR	NR	Cranial breadth and head circumference decreased	NR	Cranial breadth and head circumference decreased
Jimenez and Barrone [19]	USA	HIC	Case series	High	Low	9	NR	6.5	Sunrise technique	3.1	159	117	<5 days for all	0 (0)	NR	Symmetry achieved	18–44	Symmetry achieved with comparable blood loss and operative time
Kaiser [20]	USA	HIC	Case series	High	Low	7 (0/7)	NR	8	Extended craniectomy	NR	NR	NR	NR	NR	NR	Most patients had cranial remodeling	7	Cranial remodeling can be achieved
Kamel et al. [21]	USA	HIC	Case series	High	Low	5 (5/0)	NR	22.9	Posterior rotational cranial-flap technique using internal distraction osteogenesis	1.75	NR	NR	3.4	1 (20)	NR	Corrected craniofacial contour	26.9	Effective in maximizing bone flap viability and limiting postoperative relapse in these patients
Komuro et al. [22]	Japan	HIC	Case report	Critical	Very low	1 (1/0)	NR	8	Distraction osteogenesis combined with a barrel stave osteotomy	NR	NR	NR	NR	NR	NR	Symmetry achieved	12	Symmetry achieved
Mittermiller et al. [23]	USA	HIC	Case series	High	Low	7 (7/0)	NR	9.4	Endoscopic spring-mediated distraction	1	NR	NR	1	0 (0)	NR	Angle of the cranial base improved 5.8°	13	Endoscopic spring-mediated distraction is safe and effective

Table 2 (continued)

Paper	Country	Income status	Study design	Risk of bias	Quality grade	Isolated lambdoid craniostosis undergoing surgery (unilateral/bilateral), n	Mean age at diagnosis, months	Mean age at surgery, months	Type of surgery	Mean operative time, h	Mean blood loss, mL	Mean postoperative blood transfusion, units, mL	Hospitalization length, days	Major complications, N (%)	Reoperation, N (%)	Head contour	Mean length of follow-up, months	Key findings
Muakkassa et al. [24]	USA	HIC	Case series	High	Low	65	7	NR	Craniectomy	NR	NR	NR	NR	NR	NR	NR	NR	61 (96%) had excellent or good results All 4 patients treated before 3 months of age had excellent results All 4 patients with poor results were operated on after 6 months of age Effective and safe method
Park and Yoon [25]	Korea	HIC	Case series	High	Low	2 (2/0)	NR	10.5	Trans-sutural distraction osteogenesis	NR	NR	NR	NR	NR	NR	NR	NR	Effective and safe method
Park and Yoon [26]	Korea	HIC	Case series	High	Low	26 (26/0)	NR	10.8	Trans-sutural distraction osteogenesis	1.15	NR	22	NR	NR	NR	NR	41.9	Effective and safe method Children with lambdoid or sagittal craniostosis had most favorable outcomes
Persing et al. [27]	USA	HIC	Case series	High	Low	1 (1/0)	2	NR	Bilateral parieto-occipital cranioplasty	NR	NR	NR	NR	0 (0)	NR	Symmetric frontal and parieto-occipital regions and calvarial height is normal	12	Effective and safe
Pople et al. [28]	USA	HIC	Case series	High	Low	9 (7/2)	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	Improved or stabilized plagiocephaly
Rattani et al. [29]	USA	HIC	Retrospective cohort	Moderate	Moderate	19 (19/0)	11.12 for CVR 3.27 for ES	11.68 for CVR 3.88 for ES	Cranial vault remodeling versus endoscopic suturectomy	3.18 for CVR 0.90 for ES	210 for CVR 2.5 for ES	1-patient ES 3-patient CVR	3.17 for CVR 1.23 for ES	1 (5.3)	0 (0)	Improved with no difference between CVR and ES	54.63±5.59 for CVR 38.29±28.92 for ES	ES provides favorable outcomes relative to CVR
Rodriguez-Feo et al. [30]	USA	HIC	Case series	High	Low	21 (18/3)	8	10.75	Tongue and groove technique	3.32	388 mL	NR	6	0 (0)	5 (23.8)	Improved	54	"Tongue and groove" technique has good esthetic outcomes
Salehi et al. [31]	USA	HIC	Case series	High	Low	5	NR	5.9	Endoscopic strip craniectomy	NR	NR	NR	NR	NR	NR	NR	7.4	20% rate of neosuture development
Sato and Mitsukawa [32]	USA	HIC	Case series	High	Low	4 (4/0)	NR	24	Occipital enlargement	1.47	NR	0	NR	0 (0)	NR	Improved	61	Clinical and radiologic improvement in all cases

Table 2 (continued)

Paper	Country	Income status	Study design	Risk of bias	Quality grade	Isolated lambdoid cranio-synostosis undergoing surgery (unilateral/bilateral), n	Mean age at diagnosis, months	Mean age at surgery, months	Type of surgery	Mean operative time, h	Mean blood loss, mL	Mean postoperative blood transfusion, units, mL	Hospitalization length, days	Major complications, N (%)	Reoperation, N (%)	Head contour	Mean length of follow-up, months	Key findings
Scott et al. [33]	USA	HIC	Case series	High	Low	4 (4/0)	NR	NR	Posterior cranial vault remodeling and suboccipital decompression	NR	NR	NR	NR	NR	1 (25.0)	Improved posterior fossa size Chiari malformation resolved, improved, or stable	NR	Low complication rate and appears to be effective
Sgouros et al. [34]	USA	HIC	Case series	High	Low	3 (3/0)	NR	13.67	Circumferential-occipital craniectomy or bilateral bone flap craniotomy	NR	NR	NR	NR	NR	0 (0)	2 good outcome, 1 residual occipital deformity	NR	Effective and safe method
Sloan et al. [35]	USA	HIC	Case series	High	Low	37 (30/7)	NR	3.29	NR	NR	NR	NR	NR	NR	4 (10.8)	Improved for most patients	NR	Most had a good-excellent outcome
Smartt et al. [36]	USA	HIC	Case series	High	Low	6 (6/0)	5.2	8.3	Posterior vault switch cranioplasty	NR	NR	NR	4	0 (0)	NR	Dysmorphism improved but contralateral hemifacial deficiency persisted	84	Posterior vault switch cranioplasty is effective when performed in the first year of life
Teichgraber et al. [37]	USA	HIC	Retrospective cohort	Moderate	Moderate	NR	NR	NR	Extended strip craniectomy	NR	NR	NR	NR	NR	1 (20) in open minor group	NR	NR	No difference in reoperations between microscopic and open minor approach
Thaller et al. [38]	USA	HIC	Case series	High	Low	19 (19/0)	NR	NR	Calvarial reshaping with occipital rotation flap	NR	NR	NR	NR	0 (0)	2 (10.5)	Calvarial shape and ipsilateral ear position improved	NR	Safe and effective in improving head contour
Thatikunta et al. [39]	USA	HIC	Case series	High	Low	11 (6/5)	NR	27	Posterior cranial vault remodeling and distraction osteogenesis	2.68	86	65.27	NR	0 (0)	NR	Posterior fossa volume increased by 38.7% and foramen magnum area increased by 26.9%	NR	Highest volumetric increases are seen in unilateral lambdoid synostosis Extending osteotomy beyond the asterion is not necessary for maximal posterior vault volumetric gain
Vander Kolk et al. [40]	USA	HIC	Case series	High	Low	12 (8/4)	NR	12	Occipital bar advancement and internal osteotomies	NR	NR	NR	NR	0 (0)	NR	All had improvement in skull shape	NR	Placement of distractor in the inferior third of the suture leads to maximal posterior vault volumetric gains Good-excellent results in all patients

Table 2 (continued)

Paper	Country	Income status	Study design	Risk of bias	Quality grade	Isolated lambdoid cranio-synostosis undergoing surgery (unilateral/bilateral), n	Mean age at diagnosis, months	Mean age at surgery, months	Type of surgery	Mean operative time, h	Mean blood loss, mL	Mean postoperative blood transfusion, units, mL	Hospitalization length, days	Major complications, N (%)	Reoperation, N (%)	Head contour	Mean length of follow-up, months	Key findings
Vinchon et al. [41]	France	HIC	Retrospective cohort	Moderate	Moderate	6	NR	15.7	Multiple	NR	NR	NR	NR	NR	NR	NR	NR	Isolated LS can be managed conservatively
Weimin et al. [42]	China	UMIC	Case series	High	Low	6 (6/0)	NR	NR	Distraction osteogenesis with Z-shaped osteotomy	NR	NR	NR	NR	0 (0)	NR	All had aesthetically improved results	24	Effective method
Wilbrand et al. [43]	Germany	HIC	Case series	High	Low	1	NR	NR	NR	NR	NR	NR	NR	NR	NR	Posterior symmetry ratio improved from 0.733 to 0.808 postoperatively	NR	Can improve cranial asymmetry
Wilbrand et al. [44]	Germany	HIC	Case series	High	Low	3 (3/0)	NR	NR	Unilateral approach using a square meander-shaped craniotomy and subsequent inclination	2.63	150	NR	4	NR	NR	Cranial vault asymmetry index, posterior symmetry ratio, and posterior/anterior skull volume ratio were improved but not normalized completely	NR	Can be used for well-selected cases
Zoller et al. [45]	Germany	HIC	Case series	High	Low	21 (14/7)	NR	13.1	Occipital advancement	3.14	640.5	NR	NR	1 (4.8)	NR	Improved	NR	Remodeling leads to a well-proportioned skull shape, and posterior advancement leads to an increase in intracranial volume
Zubovic et al. [46]	USA	HIC	Case series	High	Low	12 (12/0)	10.8 for open	12.8 in open	Open cranial vault remodeling, endoscopic suturectomy	5 for open	350 in open	8 open patients received	NR	NR	NR	Posterior fossa deflection, petrous ridge asymmetry, mastoid cant, and anteroinferior ipsilateral ear displacement were not fully corrected by open reconstruction or endoscopic-assisted suturectomy	18.6 for open	Patients treated with both open and endoscopic repair of lambdoid synostosis show persistent cranial base and posterior cranial vault asymmetry
							5.1 for endoscopic-plc	5.5 in endoscopic-plc		1 for endoscopic-plc	15 in endoscopic-plc	0 endoscopic-patients received				Posterior cranial vault asymmetry improved in both groups	13 for endoscopic-assisted suturectomy	Results of endoscopic-assisted suturectomy with postoperative molding helmet therapy are similar to those of open calvarial vault reconstruction

Table 3. Categorical display of genetic mutations, inheritance patterns, and phenotypic features associated with syndromes that display lambdoid craniosynostosis

Syndrome	Mutation	Inheritance	Other features
Crouzon	<i>FGFR2, FGFR3</i>	Autosomal dominant	Midface hypoplasia, exophthalmos, cervical vertebral fusion, hearing loss, hydrocephalus
Sathre-Chotzen	<i>TWIST1, FGFR2</i>	Autosomal dominant	Parietal foramina, syndactyly, heart defects, hydrocephalus
Antley-Bixler	<i>FGFR2</i>	Autosomal recessive	Midface hypoplasia, choanal atresia, joint contractures, radiohumeral synostosis, hydrocephalus, dysmorphic facies, renal malformations, gynecologic malformations, syndactyly, arachnodactyly

FGF, fibroblast growth factor.

sular membrane around the developing brain [54–56]. The outer mesenchymal layer is then formed by intra-membranous ossification, which is dependent on the direction of forces applied by the developing brain. The developing brain is also surrounded by dural fibers, which are attached to sutures at sites of approximation of membranous bones and become sites of bony expansion [57–63]. This process is controlled by deposition of osteoid, bone remodeling, and displacement by the expanding brain, and sutural fusion is regulated by the underlying dura. Though the pathogenesis of craniosynostosis is believed to be multifactorial, there are a handful of mutations in molecular pathways and transcription factors that are implicated in disease development [63–68]. The most ubiquitous is fibroblast growth factor (FGF) [53, 60, 63, 69]. A gain-of-function mutation is implicated in several syndromic causes of craniosynostosis including Crouzon, Saethre-Chotzen, and Antler-Bixley syndromes (Table 3) [53, 69–73]. The FGF receptor is a tyrosine kinase and, among other functions, regulates osteoblast differentiation and maturation. Transforming growth factor beta has also been shown to be involved in craniosynostosis [74, 75]. The transcription factor TWIST is a regulator of FGFR1, FGFR2, FGFR3, and the osteogenic transcription factor Runx2 and has been associated with the Saethre-Chotzen syndrome [53, 64, 72, 76–78]. The transcription factor MSX2 has also been shown to cause skull ossification defects in a loss of function mutation [79, 80]. In addition to these molecules, there are also aberrant mechanical signals from the developing brain and mesenchymal cells that migrate to sutures that are involved in the development of craniosynostosis. Additionally, there are numerous environmental factors that are implicated in the development of craniosynostosis, including intra-

uterine head restraint, abnormal fetal positioning, oligo-hydramnios, teratogen (such as tobacco and valproate) exposure, prematurity, macrosomia, nulliparity, plurality, low birth weight, and shunted hydrocephalus [49, 81–85]. These causes are believed to account for 80% of all cases. Genetic mutations are often single mutations. Chromosomal abnormalities and syndromic conditions lend to the classification of syndromic and nonsyndromic types of craniosynostosis [53, 60, 86, 87]. Most syndromes are autosomal dominant, but up to 50% of cases arise de novo [69, 88]. Incomplete penetrance and variable expressivity complicate this picture [72, 89]. There have also been a small handful of reported cases where isolated lambdoid craniosynostosis was passed from father to son [3]. In the developing embryo, Virchow's law provides a basis for the occurrence of craniosynostosis: premature fusion leads to excessive growth perpendicular to patent suture along the path of least resistance [90–92].

Diagnosis

Craniosynostosis is a clinical diagnosis that is usually made in the first year of life [91]. The diagnosis begins with thorough prenatal and family histories. Careful attention must be paid to the child's progress with developmental milestones. Though in the normal adult the lambdoid suture usually fuses around 26 years of age, in children with craniosynostosis the average age is around 9.6 months [5]. Craniosynostosis can be divided into syndromic versus nonsyndromic, simple (single suture) versus complex (multiple suture), and primary (disorders of intramembranous ossification) and secondary (defective ossification related to hematologic or metabolic disorders, microcephaly from failed brain growth, shunt placement in children with hydrocephalus) subtypes [58]. In-

Pathology	Posterior view	Oblique view	Anterior view	Lateral view
Posterior plagiocephaly
Lambdoid craniosynostosis

Fig. 2. Similarities and differences between posterior plagiocephaly and lambdoid craniosynostosis from posterior, oblique, anterior, and lateral views (Courtesy of Roger Avila).

terestingly, the premature fusion of the asterion or menodosal sutures can cause a lambdoid craniosynostosis phenotype with open lambdoid sutures – an important caveat for clinicians to keep in mind [53, 93, 94].

Diagnosis based on the physical exam can be challenging, as lambdoid craniosynostosis can be difficult to differentiate from positional plagiocephaly [92, 95]. The visual similarities and differences of these 2 conditions are displayed in Figure 2. Children with lambdoid craniosynostosis have trapezoid-shaped skulls, sutural ridging, changes in ear position, downward tilt of the skull base to the ipsilateral side, prominence of the ipsilateral mastoid process which draws the ear inferiorly, and compensatory bossing of the contralateral parietal bone [96, 97]. Classically, posterior movement of the ear was said to distinguish lambdoid craniosynostosis, but recent studies have demonstrated that the inferior movement of the ear is a more accurate predictor [24]. Of note, these findings can be variable from patient to patient, making accurate diagnosis all the more challenging. Other features can include hyper- or hypotelorism, hypoplasia of the mid-face, abnormal size and shape of the ears, prominent scalp blood vessels, altered fontanel morphology, and signs of intracranial hypertension such as papilledema or sun-downing [88, 93, 97]. Children with syndromic cranio-

synostosis (SC) also display features such as syndactyly, hearing loss, and other deficits [98, 99].

The sequelae of lambdoid craniosynostosis, aside from other syndromic abnormalities, are primarily related to elevated ICP [54, 58, 88, 96, 100–104]. Sensory, respiratory, and neurological function may result, and up to 37% of children with lambdoid craniosynostosis may have intellectual disability. It is thought that learning disabilities, especially in children with SC, result from restricted brain growth, hydrocephalus, prematurity, and secondary deformations caused by high ICP and large sutures [105].

Imaging

Proper selection of imaging modalities is necessary to properly categorize and confirm diagnosis of lambdoid craniosynostosis, rule out similar conditions, and guide the surgeon toward the appropriate surgical intervention. While other craniosynostoses, such as multisuture non-SC and SCs, can be largely diagnosed from physical exam and neurological sequelae, lambdoid craniosynostosis is rarer and suture patency is more difficult to observe [106]. CT is the gold standard for imaging and confirmation of true synostoses due to its accuracy and ability to further evaluate structural abnormalities [91]. Other imaging modalities, such as magnetic resonance imaging (MRI), ultrasound,

and plain radiography, can be used but show less precision in the suture structure. Radiographic presentation can display unilateral or bilateral fusion of the lambdoid sutures, with bilateral fusion present in only 15% of cases of lambdoid craniosynostosis [107]. Unilateral lambdoid synostosis displays a bony prominence along the sutural line, ipsilateral occipitoparietal flattening, and contralateral occipitoparietal and frontal bossing [66]. Bilateral lambdoid craniosynostosis displays bilateral occipitoparietal flattening, underdevelopment of the posterior fossa, and turricephalic deformities [106, 108]. An even rarer presentation, known as “Mercedes Benz” Pattern craniosynostosis, shows an additional sagittal suture fusion in addition to bilateral lambdoid craniosynostosis [109]. Chiari malformations are also commonly seen in patients with bilateral lambdoid craniosynostosis, mainly due to overcrowding of the posterior cranial fossa [110]. The goal of postoperative imaging is to evaluate the maintenance of head shape and monitor any appearance of new symptoms. Postoperative longitudinal imaging studies are not indicated and have been replaced by nonradiological exams such as 3D laser, morphometric assessment, and smartphone-based photogrammetry [106]. Thorough radiographic analysis of patients with lambdoid craniosynostosis is necessary for optimal outcomes and management.

Surgical Management

The surgical treatment of craniosynostosis has evolved significantly over time. The unique challenges of caring for patients at such a young age, which carries a higher risk profile, have led to multiple evolutions in treatment for this condition. Initially, open strip craniectomy was the treatment of choice. While patients initially fared well, sutures would often reclose leading to unsatisfactory cosmetic results and recurrence of symptoms in more severe cases. The improvement in operative technique, technology available within the operating room, and improvements in anesthesia administration dovetailed closely with the emergence of craniofacial surgery as a discipline unto itself. What was once solely within the purview of the neurosurgeon grew to include plastic surgeons as well as many other multi-disciplinary contributors to ensure the best patient care. In modern practice at most large academic institutions, these operations are taken on as full cranial vault reconstructions with multiple teams. This entails not only removing the pathologic segment of skull as done historically but also more closely modeling the skull to normal physiology to allow for appropriate growth of the skull and brain without risk of increasing ICP and finally to achieve cosmesis.

An important consideration in undertaking this operation is the optimal time point for surgery. This is a debated topic within the field, as the balance of risks and benefits changes with time. Intervening when the patient is younger (e.g., 2–6 months) provides a simpler operation because the bone is softer and more malleable and can be more easily removed. Endoscopic suturectomy can be used during these cases [111]. However, younger age confers a larger anesthetic risk with the potential for lasting effects, and the treatment benefits may not be as durable for endoscopic procedures, even requiring helmet orthoses for significant periods postoperatively to maintain positioning. Waiting for the patient to mature further (6–12 months) allows for a more definitive correction of the deformity and more directly achieved cosmesis via contouring [112]. However, the bone has become further mineralized and is more difficult to manipulate while also causing more bleeding and a greater rate of side effects.

Lambdoid synostosis provides its own unique challenges in surgical management due to its rarity and intimate anatomic relationship with many crucial neurovascular structures. A thorough physical exam is critical, as lambdoid synostosis can be easily mistaken for positional plagiocephaly, and vice versa. As their treatments are opposite, consisting of operative correction for lambdoid craniosynostosis or positional changes or orthoses for positional plagiocephaly, the diagnosis should be verified prior to any attempted surgical intervention. Physical exam findings can distinguish the 2 conditions. While both have occipital flattening, the position of the mastoid and the ear is closely observed. In true lambdoid synostosis, the ipsilateral mastoid is elongated, and the ipsilateral ear is inferiorly displaced. Contralateral bossing of the frontal and parietal bones can be observed as well, secondary to brain expansion opposite the fused suture. Positional plagiocephaly does not typically present with these findings [113]. Additionally, the skull base itself is slightly tilted, resulting in a trapezoid shape in the head when viewed from above. Positional plagiocephaly involves a parallelogram shape when viewed from the same vantage point. CT scan of the head is a valuable tool to confirm a suspected diagnosis and it is highly specific for true synostosis. Side effects of ionizing radiation (tumorigenesis, developmental delay) in this age-group should always be considered prior to obtaining imaging. As part of the preoperative planning, obtaining an MRI of the head can be useful. Lambdoid synostosis can result in deformities to the posterior fossa. An increased proportion of these patients may concomitantly carry a Chiari malformation, especially if the synostosis is bilateral [114].

Table 4. Comparison of technical attributes, benefits, and considerations between open cranial vault reconstruction and endoscopic suturectomy to treat lambdoid craniosynostosis

Technique	Technical attributes	Benefits	Drawbacks	Considerations
Open cranial vault reconstruction	<p>Patient in prone position</p> <p>Zigzag/bicoronal incision made posterior to coronal suture, followed by periosteal dissection from inferior ipsilateral transverse sinus to contralateral transverse sinus</p> <p>Removal of parietal and occipital bones with parietal strip used to overcorrect flat side</p> <p>Barrel-Stave osteotomies used to expand cranium</p>	<p>No sacrifice in cosmetic outcomes relative to endoscopic approaches</p>	<p>Greater blood loss</p> <p>Increased operative time</p> <p>Increased anesthesia time</p> <p>Increased amount of days spent in the ICU and the hospital</p> <p>Increased transfusion rates</p>	<p>Surgical site infection</p> <p>Meningitis</p> <p>Dural tears</p> <p>Cerebrospinal fluid leak</p> <p>Blood loss</p> <p>Air emboli secondary to injured dural sinuses</p> <p>Nosocomial infection during recovery</p> <p>Comfortability of surgeon with technique</p>
Endoscopic suturectomy [112]	<p>Patient in prone position</p> <p>Two incisions made perpendicular to fused suture, one at midline of lambdoid-sagittal junction, one at mastoid bulge</p> <p>Burr hole placed at each incision, enlarged via Kerrison rongeur</p> <p>Burr holes connected via endoscope followed by use of bone-cutting scissors to separate parietal and occipital bones</p>	<p>Decreased operative time</p> <p>Decreased anesthesia time</p> <p>Lower estimated blood loss</p> <p>Decreased amount of days spent in ICU and total hospital days</p> <p>Lower transfusion rates</p>	<p>Suboptimal aesthetic correction compared with open reconstruction</p> <p>Many recommend external cranial orthoses</p>	
Spring-assisted distraction Osteogenesis [113, 114]	<p>Patient in prone position</p> <p>Zigzag/bicoronal incision made posterior to coronal suture, followed by periosteal dissection from inferior ipsilateral transverse sinus to contralateral transverse sinus</p> <p>Horizontal osteotomy of fused lambdoid suture is performed</p> <p>1 cm of bone strip is left intact to serve as a hinge and to prevent inward collapse of bone flap</p> <p>4–6 springs are placed within the osteotomy defect which expands slowly over time</p> <p>Springs are removed after an average of 2–3 months</p>	<p>Decreased operative time</p> <p>Decreased anesthesia time</p> <p>Lower estimated blood loss</p> <p>Increased volume and circumference of the cranium while minimizing dural dissection</p>	<p>Requires a second procedure to remove indwelling springs</p> <p>Suboptimal aesthetic correction compared with open reconstruction</p>	

An MRI allows identification of this preoperatively, allowing for modification of the treatment approach or management of resultant hydrocephalus. MRI also localizes important vascular structures such as the torcula and the transverse venous sinuses and their relation to the suture.

The 3 main surgical approaches are open cranial vault reconstruction, endoscopic suturectomy, and spring-assisted distraction osteogenesis [112–115]. A comparison of these techniques, along with their technical attributes, benefits, and considerations, is summarized in Table 4. In open reconstruction, the patient is positioned prone, and a coronal zigzag or bicoronal incision is made slightly posterior to the coronal suture. Periosteal dissection is then carried out inferiorly below the level of the transverse sinus ipsilaterally and posteriorly to the level of the transverse sinus on the contralateral side. The parietal and occipital bones are then removed bilaterally; this can be done en-bloc or keeping a central strip of bone in place. Dura must be meticulously dissected from the bone prior to this step, as the venous sinuses may be embedded within the bone. A strip of parietal skull from the craniectomy is taken, often from the vertex of the skull in the coronal plane, and rotated inferiorly 180° to be placed at the inferior aspect of the craniectomy to ensure overcorrection of the flat side. The bones removed are contoured *ex vivo* and replaced, and fragments are connected and distracted using absorbable plates with screws and/or wires. The correction of the flat side may also be achieved by separating the occipital bones and switching the right and left sides in cranioplasty to provide increased volume to the affected side. Barrel-stave osteotomies are also made to expand the cranium [115]. In endoscopic treatment, a patient is also positioned prone, and 2 incisions are made perpendicular to the fused suture: one near the junction of the lambdoid and sagittal suture at the midline and one over the mastoid bulge. A burr hole is placed at each location over the suture and enlarged with a Kerrison rongeur. The holes are then connected via the endoscope, and after dura is freely dissected from bone, bone-cutting scissors are used to completely free the parietal bone from the occipital bone [114].

Spring-assisted distraction osteogenesis has been shown to increase the AP length of the skull and total skull circumference [113]. An osteotomy is performed with a 1-cm remnant bone strip left to prevent inward collapse. The bone flap remains attached to the lambdoid suture. Depending on age, 4–6 springs can be placed within the osteotomy defect and can expand over time, with springs being left in for an average of 76 days [113].

Surgical Outcomes and Postoperative Considerations

While the treatment of craniosynostosis has become safer over time, there are many important intraoperative and postoperative considerations. These depend on the type of operation, namely open cranial vault reconstruction versus endoscopic correction. In a retrospective series of 19 patients surgically treated with isolated lambdoid synostosis, endoscopic correction has a significantly lower rate of operative time, anesthesia time, estimated blood loss, intensive care unit days, and hospital days when compared with open correction [114]. Additionally, surgical site infection, meningitis, dural tear, cerebrospinal fluid leak, and, perhaps most importantly, blood loss are acute complications that should be assessed for. Due to the smaller circulating blood volume in infants and the high degree of bony bleeding for open reconstruction, many surgical groups have instituted protocols to decrease blood loss and transfusion rate, such as preoperative erythropoietin administration or intraoperative blood cycling [109, 116]. These complications can ultimately lead to severe anemia and end-organ injury. Lastly, a rare but devastating complication is migration of an air embolus through an injured dural sinus, leading to massive cerebral infarct, given that the lambdoid suture is anatomically intimate with the transverse sinus, torcula, and the sagittal sinus. This can be monitored for intraoperatively with precordial doppler ultrasound [92].

Initially, many centers keep the patient in the intensive care unit for closer monitoring of lab values and to ensure that critical anemia does not develop, though there is some debate whether routine ICU admission is required [117]. If the endoscopic procedure is performed, the child often wears a helmet-molding orthosis for 4–6 months after postoperative swelling has resolved to ensure the absence of refusion and shift the free bone flaps into an appropriate position as the brain expands. Late complications include contour irregularities, failure of reossification, and refusion of suture, all of which may require reoperation. Many studies have shown that endoscopic correction can achieve comparable outcomes to cranial vault reconstruction in lambdoid synostosis [114, 118]. It is important to continue to follow these patients longitudinally, as these complications can present years after surgery.

Conclusion

Lambdoid craniosynostosis is a rare variant of craniosynostosis that can lead to developmental delay, facial abnormality, and neurological dysfunction. Surgical correc-

tion with postoperative helmet orthosis is the preferred method of treatment, with encouraging long-term results. Clinicians must be able to recognize isolated lambdoid craniosynostosis based upon clinical and imaging characteristics and provide timely surgical management to avoid cosmetic and neurological deficits.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Taylor Reardon suggested and developed the idea and contributed to the Abstract, Introduction, and Radiologic Presentation sections. Dr. Brian Fiani provided mentorship and document editing and contributed to the Conclusion section. Dr. Jacob Kosarchuk provided mentorship and contributed to the Demographics, Embryological Development, Pathogenesis, and Clinical Presentation and Neurological Sequelae sections. Dr. Anthony Parisi provided mentorship and contributed to the Surgical Techniques and Surgical Outcomes and Postoperative Considerations sections. Nathan A. Shlobin conducted the systematic review and contributed to the Methods and Results sections. All the authors fulfilled the ICMJE Criteria for Authorship.

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