Bone Regrowth After Chiari I Decompression in Children with Syndromic Craniosynostosis

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Abstract

Objective:
Chiari malformation type I (CM-I) is common in children with syndromic craniosynostosis. While posterior fossa decompression is generally effective, reoperation rates remain variable, and rare failure mechanisms are poorly characterized. We aimed to analyze reoperation causes in pediatric CM-I with a focus on restenosis due to bone regrowth.
Methods:
We retrospectively reviewed all pediatric patients (<18 years) surgically treated for CM-I at a single tertiary center between 2008 and 2023. Demographic data, syndromic status, surgical technique, imaging findings, and revision surgeries were analyzed.
Results:
Thirty-seven patients were included (13 syndromic, 24 non-syndromic). Seven patients (19%) required reoperation. Restenosis due to solid bone regrowth occurred in six patients and was observed exclusively in children with syndromic craniosynostosis, all with pansynostosis. Five of six had undergone prior ventriculoperitoneal shunting.
Conclusion:
Bone regrowth represents a distinct, delayed cause of restenosis after CM-I decompression in syndromic craniosynostosis, warranting prolonged risk-adapted follow-up.

Introduction

Chiari malformation type I (CM-I) is characterized by caudal displacement of the cerebellar tonsils and may lead to brainstem compression, syringomyelia, and neurological deficits. In pediatric patients, CM-I is frequently associated with syndromic craniosynostosis due to posterior fossa hypoplasia.
Posterior fossa decompression is the standard surgical treatment; however, reported reoperation rates range widely. Most revisions are attributed to CSF-related complications or insufficient decompression. Restenosis caused by true bone regrowth is rarely reported, with only isolated cases described in the literature. Data on incidence, risk factors, and clinical relevance of this phenomenon remain scarce, particularly in syndromic pediatric populations.

Methods and Materials

A retrospective analysis was conducted of all pediatric patients surgically treated for CM-I between 2008 and 2023. Inclusion criteria comprised patients under 18 years with CM-I; CM-II and adult patients were excluded. Data collected included demographic variables, syndromic diagnosis, clinical presentation, presence of syringomyelia or hydrocephalus, operative technique, and postoperative course. Surgical management consisted of foramen magnum decompression with or without C1 laminectomy, dural opening, tonsillar reduction, and expansion duraplasty, guided by intraoperative ultrasound. Routine postoperative MRI was performed at three months and annually thereafter.

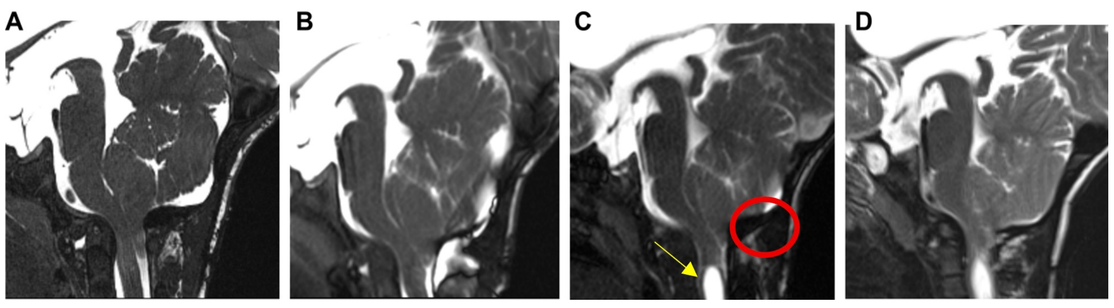

Fig. 1 A: The preoperative imaging of a child diagnosed with Pfeiffer syndrome revealed stenosis of the craniocervical junction, accompanied by the herniation of the cerebellar tonsils. B: After decompression, with resection of the lamina of the first cervical vertebra and partial resection of the cerebellar tonsils, the foramen magnum and perimedullary spaces were widened. C: Routine imaging 3 years postoperative presented suboccipital reossification (circle) causing restenosis and new syringomyelia (arrow). Hence a subsequent decompression of the foramen magnum was conducted. D: The postoperative imaging demonstrates the osseous defect up to the anterior border of the foramen magnum.

Results

A total of 37 pediatric patients underwent surgical treatment for CM-I. Thirteen patients had syndromic craniosynostosis, most commonly Crouzon syndrome. Syringomyelia was present in 51%, and hydrocephalus requiring CSF diversion in eight patients.
Seven patients (19%) required revision surgery at a mean of 3.0 years (range 9 months–8 years). In six patients, restenosis was caused by solid bone regrowth around the foramen magnum, with partial regrowth of the C1 arch in two cases. All bone regrowth cases occurred exclusively in syndromic CM-I patients. Bone regrowth occurred irrespective of the initial decompression strategy.

Discussion

This series represents the largest reported cohort of pediatric CM-I patients developing restenosis due to solid bone regrowth. Strikingly, bone regrowth occurred exclusively in patients with syndromic craniosynostosis and pansynostosis.
Pediatric cranial osteogenesis is strongly influenced by dural signaling, particularly in early childhood. Gain-of-function mutations affecting FGFR pathways and dysregulation of osteoblast activity in syndromic craniosynostosis may predispose these patients to pathological bone regeneration. Additionally, prior CSF diversion may alter biomechanical forces at the craniocervical junction, potentially contributing to osteogenesis, even in the absence of overt over-drainage.

Conclusions

Bone regrowth causing delayed restenosis after Chiari I decompression is a rare but clinically relevant complication occurring exclusively in children with syndromic craniosynostosis. These patients represent a high-risk subgroup and should undergo structured long-term clinical and radiographic follow-up to enable early detection and timely revision surgery.

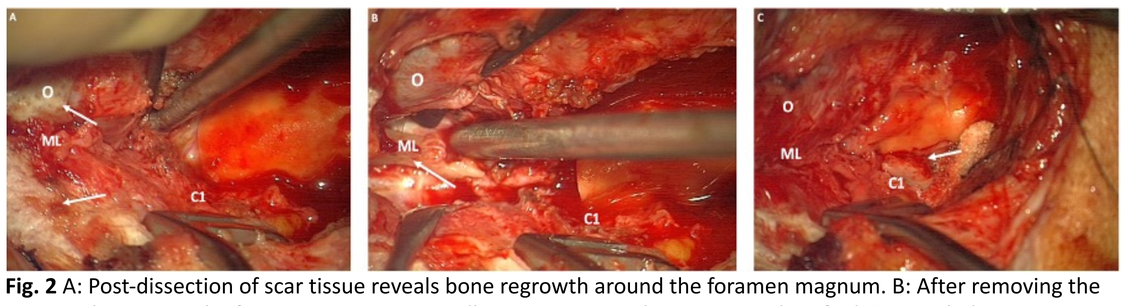

Fig. 2 A: Post-dissection of scar tissue reveals bone regrowth around the foramen magnum. B: After removing the regrown bone near the foramen magnum, a midline-compressing bone spur is identified. C: Detailed examination and clearance of scar tissue at the C1 level uncover partial regrowth of the C1 arch.
O = level of the occipital bone, ML = midline, C1 = level of the C1 arch, white arrows = areas of bone regrowth

Initial symptoms	pre-OP (n=)	post-OP improvement (n=)
Syringomyelia (51%)	19	16
Head/Neck pain (41%)	15	15
Cranial nerve deficits (16%)	6	5
Ataxia (22%)	8	7
Sleep apnea (19%)	7	5
Sensory deficit (16%)	6	6
Motor deficits (5%)	2	1

Table 1: Preoperative symptoms and postoperative improvement

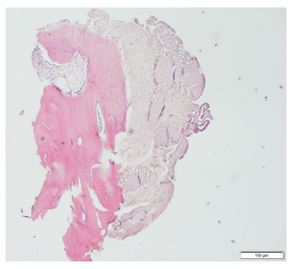

Fig. 3 Histopathological images of intraoperatively resected tissue showing hematoxylin-eosin stains with collagenous tissue (dura mater and scar tissue) to the right and bone tissue with richer eosin enhancement to the left

Initial diagnosis	VP-Shunt	Syringomyelia initially	Symptomatic initially	Initial surgical treatment	Age at 1st surgery	Syringomyelia before second surgery	Symptomatic before 2nd surgery	Bone regrowth	Revision surgery	Age at reoperation
Non-Syndromic CM-I	no	no	yes	FMD, C1	2 years	no	yes	no	FMD, DP, TR	6 years
Crouzon syndrome	yes	yes	yes	FMD, C1	3 years	yes	yes	yes	FMD, DP, TR	11 years
Crouzon syndrome	no	no	no	FMD	2 months	yes	yes	yes	FMD, DP, TR	5 years
Crouzon syndrome	yes	no	no	FMD, C1	2 years	no	yes	yes	FMD, DP, TR	2 years
Crouzon syndrome	yes	no	yes	FMD, TR, DP	5 years	yes	yes	yes	FMD, C1, DP	6 years
Pfeiffer syndrome	yes	no	yes	FMD, TR, DP	2 years	yes	no	yes	FMD	6 years
Pansynostosis	yes	no	no	FMD, TR, DP	4 years	yes	no	yes	FMD, C1, DP	6 years

Table 2: Overview of patients undergoing revision surgery, CM-I=Chiari malformation type I, VP-Shunt=ventriculoperitoneal shunt, FMD= foramen magnum decompression, C1=Laminectomy of the first cervical vertebra, TR=reduction of the cerebellar tonsils, DP= extension duraplasty

Contact

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References

1. Cinalli G et al. *Childs Nerv Syst*. 2005
2. Tam SKP et al. *Acta Neurochir*. 2021
3. Kitamura S et al. *Childs Nerv Syst*. 2022
4. Talamonti G et al. *J Clin Med*. 2023
5. Cunningham ML et al. *Orthod Craniofac Res*. 2007