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Radiological evaluation of Chiari 1 malformation: Diagnostic findings and postsurgical imaging

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ABSTRACT

Objective: Chiari 1 malformation is a congenital anomaly characterized by downward displacement of the cerebellar tonsils through the foramen magnum. Surgical decision-making in Chiari 1 largely depends on clinical symptoms and imaging findings, particularly the presence of headache and syringohydromyelia. This study aimed to evaluate the radiological characteristics and postoperative imaging changes in pediatric patients with Chiari 1 and Chiari 1.5 malformations.

Material and Methods: This retrospective study included 101 pediatric patients (age range; 1–18 years) diagnosed with Chiari 1 or Chiari 1.5 based on brain and cervical spine MRI findings between November 2022 and April 2025. Clinical and radiological data, including tonsillar descent, syringomyelia, craniovertebral junction anomalies, and basal angle measurements were analyzed. Twenty-one patients underwent posterior fossa decompression and postoperative MRIs were evaluated at 3 and 6 months. Statistical analyses were performed using chi-square, Mann–Whitney U, Wilcoxon signed-rank, and McNemar tests, with $p < 0.050$ considered significant.

Results: Among the 101 patients, 52 (51%) were female, and 49 (49%) were male (mean age; 11.1±4.9 years). Headache was significantly more frequent in operated patients (66.7% vs. 32.5%, p=0.014). Syringomyelia was also more common in the surgical group (33.3% vs. 3.8%, $p < 0.001$), and tonsillar descent was greater (14.3±5.3 mm vs. 9.5±3.6 mm, $p<0.001$). No significant differences were found between the groups regarding age, sex, basal angle, or foramen magnum diameter. Postoperative follow-up revealed regression of syrinx dimensions and clinical improvement in all operated patients.

Conclusion: Headache, syrinx formation, and greater tonsillar descent are key radiological and clinical predictors of surgical intervention in pediatric Chiari 1. While craniovertebral junction anomalies and hydrocephalus may coexist, they appear less predictive of surgical necessity. Posterior fossa decompression remains a safe and effective treatment in symptomatic pediatric patients, leading to both clinical and radiological improvement.

Keywords: Chiari 1 malformation, craniovertebral junction, pediatric MRI, posterior fossa decompression, syringomyelia

Introduction

Chiari malformation was first described in 1890 by the Austrian pathologist Hans Chiari (1). It represents a spectrum of developmental abnormalities primarily involving the posterior fossa and the craniocervical junction. To date, nine subtypes have been described in the literature (2,3). Advances in neuroimaging techniques have contributed significantly to the recognition and expansion of this classification.

For accurate diagnosis and appropriate management, it is essential to evaluate Chiari 1 not as an isolated malformation but within the context of associated deformities and related Chiari subtypes. As the primary treatment modality is surgical, the identification of subtypes, associated anomalies, and the exclusion of other potential causes of tonsillar descent are of critical importance (4).

Diagnosis is established when the cerebellar tonsils extend ≥5 mm below the foramen magnum, defined by a line between the basion and opisthion on the midsagittal plane.

Confirmation of tonsillar descent on coronal images is important for diagnostic accuracy (5).

Among the related and overlapping entities, Chiari 0 represents the presence of a syrinx without cerebellar tonsillar herniation (<5 mm below the foramen magnum). Chiari 0.5 is characterized by no tonsillar herniation (<5 mm below the foramen magnum) but with the cerebellar tonsils located ventral to the line bisecting the medulla on the midsagittal plane. Chiari 1 is defined as a caudal herniation of the cerebellar tonsils >5 mm below the foramen magnum, while Chiari 1,5 describes tonsillar herniation associated with caudal displacement of the obex below the foramen magnum (2).

In daily radiological practice, Chiari 0, 0.5, and 1.5 are less frequently used terms; the most commonly recognized entity within this spectrum is Chiari 1. The current study includes patients diagnosed with Chiari 1 and Chiari 1.5 malformations. Differentiating Chiari 1.5 from Chiari 1 is important, as herniation of the obex below the foramen magnum at the cervicomedullary junction has been associated with less favorable surgical outcomes in some reports (6).

Associated abnormalities include posterior angulation of the atlas and odontoid process, basilar invagination, platybasia, and various craniocervical junction anomalies such as vertebral fusion or segmentation defects. In Chiari İ 1, clival hypoplasia and disruption of the normal parallel alignment between Chamberlain's and McRae's lines are also frequently observed radiologic findings (7).

Syringohydromyelia is considered a complication of the disease and is thought to result from impaired CSF dynamics at the craniocervical junction (8). Approximately 30% of patients are asymptomatic (9). When symptomatic, the most common presentation is suboccipital headache induced by coughing or Valsalva maneuvers. Other manifestations include dizziness, cranial nerve dysfunction, paresthesia, and cerebellar signs (10).

The decision for surgery in Chiari 1 patients is guided by clinical presentation and imaging findings, particularly the presence of headache and syringohydromyelia (11). Neurosurgeons generally recommend surgery under these conditions (12). The most widely accepted surgical approach is posterior fossa decompression with duraplasty which typically involves suboccipital craniectomy and C1 laminectomy to achieve decompression. Postoperative radiological evaluation is essential for assessing potential complications such as cerebellar parenchymal changes and posterior fossa alterations (13-15).

This study aimed to retrospectively characterize the radiological features of Chiari 1 and evaluate postoperative imaging changes in a cohort of symptomatic and incidentally detected patients.

Materials and Methods

Brain and cervical MRI examinations performed between November 2022 and April 2025 in pediatric patients aged 0–18 years were retrospectively reviewed.

Preoperative and postoperative clinical data related to Chiari 1 were reviewed, including the presence of headache (particularly Valsalva-induced), vertigo, sensory disturbances, seizures, and syncope.

Imaging Technique

All MRI examinations were performed using a 1.5-Tesla scanner (SIGNA™ Explorer, GE Healthcare Technologies, Chicago, Illinois, USA) equipped with standard head and spine coils. The cervical spine protocol included sagittal and axial T1- and T2-weighted sequences, while the brain MRI protocol comprised axial T1-, T2-, and FLAIR-weighted, coronal T2-weighted, and 3D T1-weighted sequences. The 3D T1-weighted sequence, routinely included in the brain MRI protocol, was primarily used for detailed evaluation of the craniocervical junction. Imaging parameters included a slice thickness of 3–4 mm, an interslice gap of 0.5–1 mm, and a field of view adjusted according to patient size.

Phase-contrast cine MRI was used to assess cerebrospinal fluid (CSF) dynamics at the craniocervical junction. Imaging was performed in the mid-sagittal plane using a velocity-encoded (VENC) phase-contrast sequence gated to the cardiac cycle. Typical acquisition parameters included: repetition time (TR), echo time (TE), section thickness of 3–5 mm, and a VENC value of 5–12 cm/s to visualize bidirectional CSF pulsations. Flow-sensitive images were reconstructed to generate magnitude and phase maps for the evaluation of CSF flow voids and patency at the foramen magnum.

Imaging analysis

All imaging studies were independently reviewed by two pediatric radiologists with 15 and 8 years of experience, respectively, and discrepancies were resolved by consensus.

On brain MRI, the degree of tonsillar descent below the foramen magnum was assessed on mid-sagittal T1- or T2-weighted images; in cases of asymmetric herniation, the greater degree of descent was recorded.

Patients with tonsillar herniation <5 mm, incomplete clinical data, or herniation secondary to intracranial space-occupying lesions causing increased intracranial pressure were excluded from the study. In patients with tonsillar herniation >5 mm, the anteroposterior diameter of the foramen magnum was measured. Both Chiari 1 and Chiari 1.5 were identified, and surgical reports along with postoperative imaging studies were retrospectively examined. Follow-up MRI examinations were performed at 3 and 6 months postoperatively.

Hydrocephalus was assessed in all patients, and postoperative MRI scans were reviewed to document

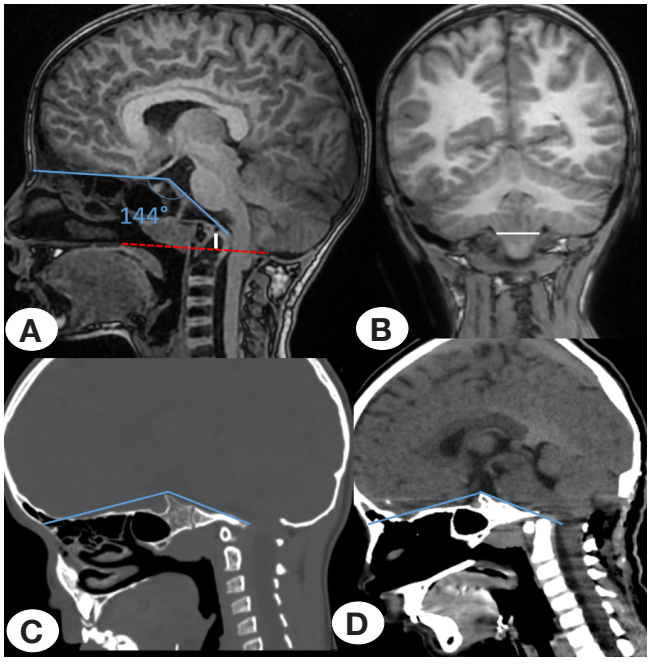

Figure 1: Associated abnormalities in Chiari 1. In a patient with Chiari 1, **A)** sagittal and **B)** coronal T1-weighted MRI, along with **C,D)** Sagittal CT demonstrates a widened basal angle measuring 144° (blue angle), consistent with platybasia. The dens projects above the Chamberlain line (dashed red line), indicating basilar invagination (white line in A). Cerebellar tonsillary herniation also shows descent of the cerebellar tonsils below the foramen magnum (white line). **D)** Following posterior fossa decompression, the basal angle remains unchanged; however, the cerebellar tonsils are restored to their normal position.

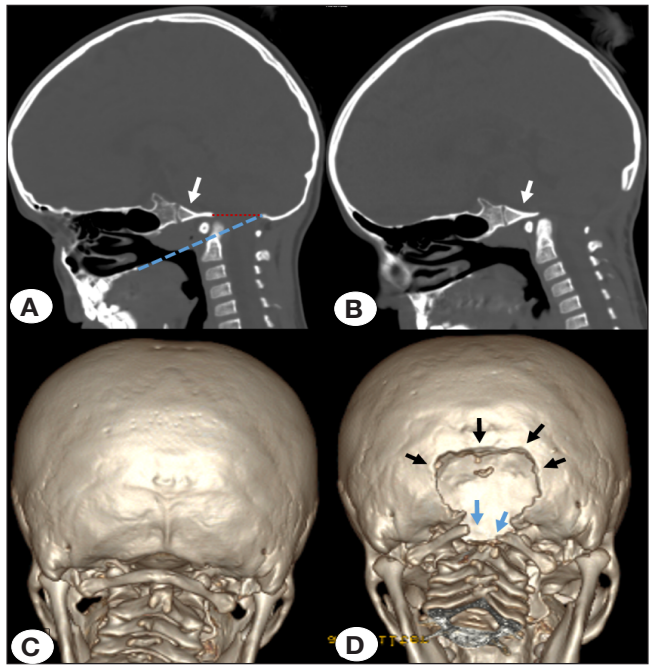

Figure 2: In a patient with Chiari I malformation, **A,B)** sagittal CT images demonstrate an abnormal angulation between the Chamberlain line (dashed blue line) and the McRae line (dashed red line). These lines are normally parallel; however, in Chiari I malformation, clival hypoplasia (white arrows) leads to loss of parallel alignment. **C,D)** preoperative and following decompression operation, 3D reformatted CT images show a post-suboccipital craniectomy defect (black arrows) and an osteotomy defect in the posterior arch of the atlas (blue arrows).

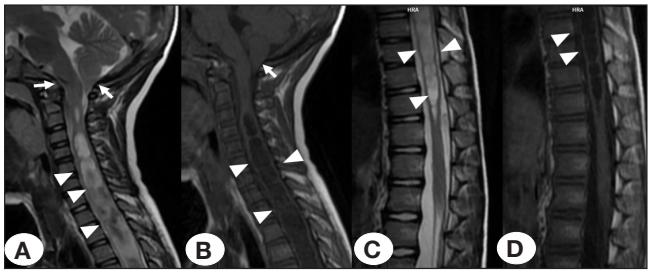

Figure 3: Syringohydromyelia. **A,B)** Sagittal T2-weighted, **C,D)** T1-weighted spinal MRI images demonstrate cerebellar tonsillar herniation with secondary foramen magnum stenosis (white arrows) and an extensive syringohydromyelia involving nearly the entire spinal cord (white arrowheads).

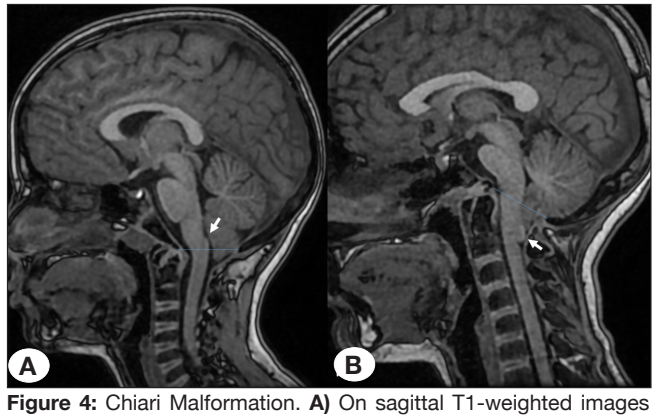

Figure 4: Chiari Malformation. **A)** On sagittal T1-weighted images cerebellar tonsillar herniation indicates a Chiari 1 when the obex (white arrow) remains above the foramen magnum (blue dashed line), **B)** a Chiari 1.5 when the obex (white arrow) is displaced caudally below the foramen magnum (blue dashed line).

ventricular changes, interval improvement or resolution, and any CSF diversion procedures performed.

Cervical spine MRI and CT—when available—were evaluated for vertebral fusion and segmentation defects and craniocervical junction abnormalities. Basal angle measurements were obtained in all patients, with pre- and postoperative comparisons performed in those who underwent surgery (Figure 1-2). Additionally, cervical MRI was reviewed for the presence, location, and dimensions of syringomyelia, and in surgically treated patients, postoperative syrinx. measurements were also recorded (Figure 3).

CSF-flow MRI was performed in patients with Chiari 1 or Chiari 1.5 malformation, as well as in those demonstrating clinical or radiologic findings suggestive of hydrocephalus, craniocervical junction anomalies, foramen magnum stenosis, or cord compression on cranial or cervical MRI (Figure 4). CSF-flow MRI was not performed in Chiari 1 patients who lacked these additional clinical or imaging indications. In patients who underwent CSF-flow MRI, the absence of the

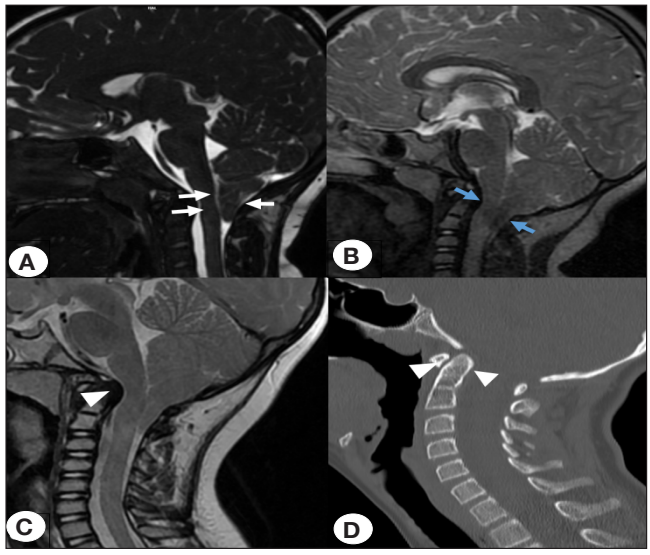

Figure 5: In a patient with Chiari 1, Sagittal FIESTA **A)** and T2-weighted **C)** MRI sequences of the craniocervical junction demonstrate marked narrowing of the foramen magnum secondary to significant cerebellar tonsillar herniation (white arrows). On the DRIVE sequence **B)**, loss of the normal cerebrospinal fluid (CSF) flow void is noted (blue arrows), with additional contribution from posterior angulation of the C2 odontoid process (arrowhead) to the foramen magnum narrowing. Similar posterior angulation of the odontoid process is also evident on the sagittal CT image **D)**.

normal CSF flow void at the level of the foramen magnum was documented (Figure 5).

Statistical analysis

Statistical analysis were performed using SPSS version 26.0 (IBM Corp.). The Kolmogorov-Smirnov test was employed to assess the normality of data distribution. Frequencies and percentages are presented for qualitative data, while for quantitative data, the mean and standard deviation are reported for variables. In the comparison between the operated and non-operated groups, categorical data were compared using the chi-square test, and quantitative variables were compared using the Mann–Whitney U test. Preoperative and postoperative syrinx dimensions and angle measurements were compared using the Wilcoxon signed-rank test, and the presence of syrinx was compared using the McNemar test. A p-value <0.050 was considered statistically significant.

Results

A total of 101 patients were included, with 21 (21%) undergoing surgical treatment for Chiari malformation and 80 (79%) managed conservatively. The majority had Chiari 1, while only 3 (3%) patients were diagnosed with Chiari 1.5; these were included in the operated group. Given the small number of Chiari 1.5 cases, subgroup statistical comparisons were not performed.

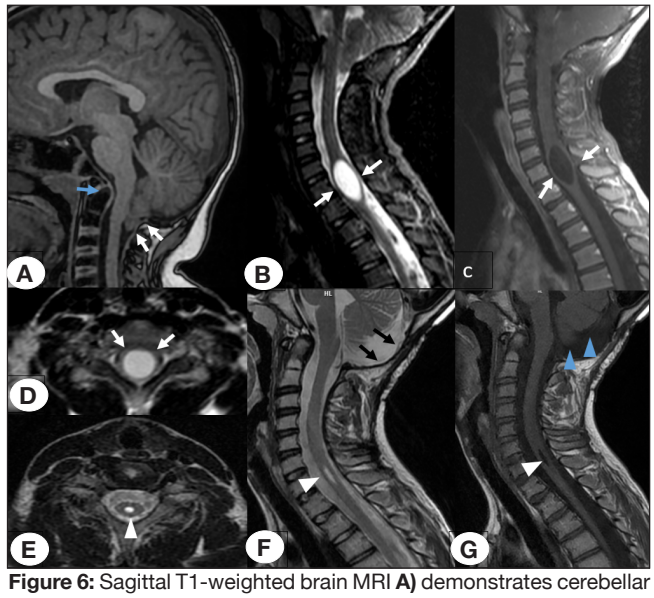

Figure 6: Sagittal T1-weighted brain MRI **A)** demonstrates cerebellar tonsillar herniation (white arrows) and posterior angulation of the C2 odontoid process (blue arrow). On preoperative cervical MRI, sagittal T2- **B)**, sagittal T1- **C)**, and axial T2-weighted **D)** images reveal a syrinx within the central spinal canal at the C7 level. In the postoperative evaluation **(E-G)**, marked regression of the syrinx is observed (arrowheads). Post-suboccipital craniectomy bone defect (black arrows), expansion of the posterior fossa, and upward repositioning and decompression of the cerebellar tonsils (blue arrowheads in f and **G)** are also noted.

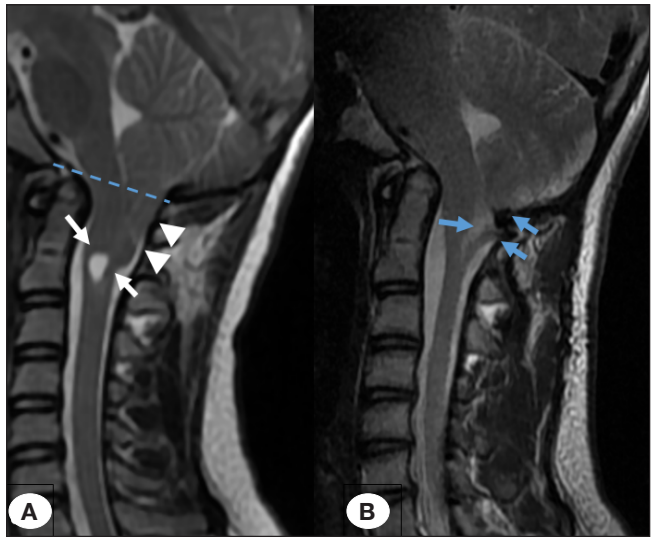

Figure 7: Sagittal T2-weighted cervical spinal MRI obtained **A)** Preoperatively and b. postoperatively demonstrates marked regression of cerebellar tonsillar herniation (arrowheads) and a residual small syrinx cavity within the spinal cord (white arrows), consistent with postoperative improvement in Chiari 1. The postoperative images also show decompression of the foramen magnum region (blue arrows) *The blue dashed line indicates the foramen magnum level.

The mean age of the cohort was 11.1±4.9 years (range; 1–18 years). There were 52 females (51%) and 49 males (49%), with no significant sex predominance observed.

Clinical symptoms attributable to Chiari 1 were absent in 44 patients (43.6%), and the diagnosis was made incidentally

Table I: Craniovertebral and spinal anomalies in patients with Chiari 1 according to surgical status

Anomaly	Operated*	Non-operated*
Posterior angulation of odontoid process	11 (52)	17 (21)
Basilar invagination	1 (5)	5 (6.2)
Platybasia	1 (5)	4 (5)
Vertebral fusion anomaly	0	2 (2.5)
Scoliosis	2 (10)	5 (6.2)
Total	21	80

***:** *n(%)*

Table II: Comparison of morphometric measurements between operated and non-operated patients

	Operated (n = 21)	Non-operated (n = 80)	p
Basal angle*	124.3±7.1	124.3±7.7	0.876
Foramen magnum diameter*	35.3±3.6	36.1±4.6	0.336
Tonsillar herniation*	14.3±5.3	9.5±3.6	<0.001

***:** *mean ±SD*

Table III: Comparison of preoperative and postoperative measurements

	Preoperative	Postoperative	p
Syrinx AP diameter (mm)*	7.25 (2-12)	3.9 (2–7)	0.043
Syrinx CC length (mm)*	55.5 (6-200)	55.1 (6-200)	0.893
Basal angle†	124.3±7.1	126.7±6.2	0.215
Presence of syrinx‡	8	7	1.000

***:** *mean (range),* **†:** *mean±SD,* **‡:** *n(%),* ***AP:** Anteriorposterior,* ***CC:** Craniocaudal*

during imaging performed for other reasons. Headache exacerbated by the Valsalva maneuver, a symptom considered characteristic of Chiari malformation, was reported in 40 patients (39.6%). Vertigo was present in 12 patients (11.9%), seizures or syncope in 1 patient each (1%), and extremity numbness in 4 patients (3.9%). Craniovertebral and spinal anomalies were observed in both operated and non-operated patients (Table I). Posterior angulation of the odontoid process was the most frequent anomaly in both group, whereas basilar invagination, platybasia, vertebral fusion anomalies, and scoliosis were less common. Two patients with scoliosis accompanied by syrinx underwent surgery. Patients without syrinx were followed up (n=5). No vertebral fusion anomalies were identified in any of the operated patients.

Comparisons between operated and non-operated Chiari 1 patients revealed several significant differences. Headache was significantly more frequent in the operated group (66.7% vs. 32.5%, p=0.014), and syrinx formation was markedly more common (33.3% vs. 3.8%, p<0.001). In addition, the mean tonsillar herniation distance was significantly greater in operated patients (14.3±5.3 mm) compared with non-operated patients (9.5±3.6 mm, p<0.001) (Table II).

Here were no statistically significant differences between the operated and non-operated groups regarding age, gender distribution, vertigo, syrinx dimensions (anteroposterior or craniocaudal), foramen magnum diameter, basal angle (p> 0.050 for all). Overall, these results suggest that headache, syrinx formation, and greater tonsillar descent are strongly associated with surgical intervention, whereas the presence of other craniovertebral anomalies appears less predictive (Table III).

Overall, clinical symptoms improved in all operated patients, and, when present, syrinx anteroposterior diameters demonstrated regression (Figure 6,7).

Hydrocephalus was identified in seven patients, six of whom underwent surgical intervention. One patient was managed with a ventriculoperitoneal (VP) shunt. The presence of hydrocephalus was significantly associated with the decision to perform posterior fossa decompression (p <0.001). All patients with hydrocephalus demonstrated a decrease in ventricular size postoperatively, and none required an additional CSF diversion procedure.

All operated patients underwent a posteroinferior occipital craniectomy combined with a laminectomy of the posterior arch of the atlas, and seven patients additionally underwent duraplasty. Clinical improvement was observed in all cases.

In one patient who underwent duraplasty with tonsillar resection, early postoperative diffusion restriction consistent with cytotoxic edema was observed in the cerebellar tonsils; this finding resolved on both the 3- and 6-month follow-up scans.

Discussion

The evaluation of the cerebellar tonsils in relation to the foramen magnum is essential in routine cranial and cervical MRI or CT assessment. Tonsillar descent <5 mm is defined as tonsillar ectopia, whereas descent >5 mm is considered tonsillar herniation, consistent with Chiari malformation. In pediatric patients with Chiari 1, cervical or whole-spine MRI is recommended to assess for associated anomalies such as syringomyelia, tethered cord, and craniocervical junction abnormalities (16). Management typically involves follow-up for ectopia or herniation <1 cm, while herniation >1 cm accompanied by symptoms, syrinx, or hydrocephalus may warrant surgical intervention (17).

In this retrospective study of 101 pediatric patients with Chiari 1 and Chiari 1.5, we observed that headache, syrinx formation, and greater tonsillar descent were significantly associated with the decision to perform surgical intervention. The majority of patients presented with Chiari 1, and only a

small subset had Chiari 1.5, consistent with previous reports highlighting the rarity of the latter in the pediatric population (18). Although Chiari 1.5 are less frequently observed, the obex in these patients is positioned below the foramen magnum, contributing to increased foramen magnum stenosis and neural compression, which may accentuate clinical manifestations. Several studies have also proposed that Chiari 1.5 represents a progressive variant of Chiari 1 (19). A hypothesis that may account for its relative rarity in the pediatric population.

Headache exacerbated by the Valsalva maneuver was the most common clinical symptom in operated patients, supporting its utility as a predictive symptom for surgical intervention. This aligns with prior studies that have identified Valsalva-induced headache as a hallmark feature of symptomatic Chiari malformation (20,21). Dizziness, parestehesias, seizures, and syncope were observed less frequently in the cohort; however, notable improvement in these symptoms was also documented following surgical decompression. Symptoms such as headache and paresthesia may provide important guidance in the differential diagnosis of other neurological disorders. Although numbness and paresthesias are often interpreted as suggestive of demyelinating disease, it is crucial to assess these manifestations within the context of Chiari 1, as they can also occur as a direct consequence of the malformation itself (2).

Syringomyelia was significantly more prevalent in the operated group, consistent with the well-established association between syrinx presence and symptomatic Chiari malformation (22,23). In our cohort, surgical decompression resulted in regression of syrinx dimensions in all patients, supporting the efficacy of posterior fossa decompression in restoring normal CSF dynamics and reducing syrinx size. In our study, postoperative follow-up imaging was conducted at 3 and 6 months. During this interval, a significant reduction in the anteroposterior diameter of the syrinx was observed; however, no appreciable decrease in the craniocaudal dimension was detected. This discrepancy may reflect the relatively short follow-up period, underscoring the need for longer-term imaging to fully assess syrinx resolution. Notably, the mean tonsillar herniation distance was greater in operated patients, corroborating previous studies that have suggested the degree of herniation as a factor influencing surgical decision-making (24,25).

Following the assessment of cerebellar tonsillar herniation, it is essential to evaluate the craniocervical junction for anomalies and potential instability. Growing evidence suggests that craniocervical instability may contribute to tonsillar herniation in at least a subset of patients with Chiari 1 (5,26,27). In our cohort, craniovertebral junction anomalies—including posterior angulation of the odontoid process, basilar invagination, and platybasia—were frequently observed. Although these anomalies did not appear to influence the decision for surgical intervention in our study, their presence remains clinically significant, as they may impact patient morbidity and long-term outcomes.

In our cohort, hydrocephalus emerged as a strong predictor for surgical intervention, aligning with previous reports identifying ventricular enlargement as a marker of disease severity in Chiari 1 (28,29). Nearly all patients with hydrocephalus underwent posterior fossa decompression, and the statistical association between hydrocephalus and the decision for surgery was highly significant ($p<0.001$). This finding supports the notion that concurrent hydrocephalus not only reflects increased intracranial pressure dynamics but also influences neurosurgical management strategies. Early recognition of hydrocephalus may therefore facilitate timely surgical planning and potentially improve postoperative outcomes.

All surgeries consisted of a posteroinferior occipital craniectomy with C1 laminectomy, and seven patients underwent duraplasty. Clinical improvement was observed in all patients, and postoperative imaging demonstrated resolution of syrinx and, in one case, transient diffusion restriction in the cerebellar tonsils that resolved by 3 months. These findings indicate that posterior fossa decompression is generally safe and effective in children, with low rates of transient complications and favorable radiological and clinical outcomes (30).

Limitations

Limitations of this study include its retrospective design and the single-center setting, which may limit generalizability. Additionally, thoracic and lumbar vertebral anomalies were not evaluated, and the small number of Chiari 1.5 patients precluded subgroup analysis. Prospective, multicenter studies with larger cohorts are needed to further delineate predictors of surgical intervention and long-term outcomes in pediatric Chiari malformations.

Conclusion

In conclusion, our findings emphasize that headache, syrinx formation, and increased tonsillar descent are key factors associated with the decision to operate in pediatric Chiari 1. While craniovertebral junction anomalies may coexist, they appear less predictive of surgical necessity. Posterior fossa decompression is effective in achieving clinical improvement and syrinx regression, supporting its continued role as the primary surgical intervention in symptomatic pediatric patients.

Ethics committee approval

This study was conducted in accordance with the Helsinki Declaration Principles. The study was approved by Etlik City Hospital (13.03.2025, reference number: 2025-124).

Contribution of the authors

Study conception and design: ŞY, BU; data collection: ŞY, ZZDÖ; analysis and interpretation of results: ŞY, HB, MEE; draft manuscript

preparation: ŞY, BU. All authors reviewed the results and approved the final version of the article.

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Conflict of interest
The authors declare that there is no conflict of interest.

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