

Prevalence of High-riding Vertebral Artery in Chiari Malformation Type-1

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ABSTRACT

Objective: The aim of this study is to analyze the patients operated on for Chiari malformation type 1 for the presence of a high-riding vertebral artery (HRVA) and to investigate the effect of this parameter on surgery.

Materials and Methods: Patients who underwent surgery for Chiari malformation type 1 between 2018 and 2020 in our clinic were retrospectively evaluated with patient files and preoperative cervical spine computed tomography (CT) imaging. A total of 15 patients were included in the study. Lateral mass internal height and isthmus height measurements were performed to investigate the presence of HRVA.

Results: One (3.3%) of 30 C2 lateral mass internal height measurements showed a high-riding vertebral artery on the right side. A high-riding vertebral artery was found in 13 of 30 C2 isthmus height measurements (43.3%). Of these, 3 were right-sided, 2 were left-sided, and 4 were bilateral. A total of 9 patients had unilateral or bilateral HRVA. None of these patients experienced any surgical morbidity related to HRVA.

Conclusion: Although HRVA was detected at a relatively high rate in CM-1 patients in our series, it did not affect the surgical outcome. We conclude that investigating the presence of HRVA in the preoperative planning of CM-1 patients could be helpful in avoiding possible complications.

Keywords: Cerebellum, chiari malformation, high-riding vertebral artery, skull base

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INTRODUCTION

Chiari malformation (CM) is one of several structural deformities of the skull base and cerebellum. It is characterized by the displacement of the cerebellar tonsils caudal to the foramen magnum. It was first named by the Austrian pathologist Hans Chiari. Chiari malformation is seen in four types, and the most common type is Chiari malformation type 1 (CM-1).^[1] Cases in which cerebellar tonsil herniation extends more than 5 mm into the spinal canal are considered CM-1. This herniation causes the brainstem and spinal cord to be compressed, and the circulation of cerebrospinal fluid (CSF) is impaired.^[2]

Many theories have been proposed regarding the etiology of CM. Incomplete posterior fossa development and the resulting structural deformities constitute one of these theories.

^[3] Another theory is that circulatory disturbance of the CSF

leads to pressure changes and causes herniation. Pressure on the brainstem and cerebellum caused by fetal hydrocephalus is also implicated. Nevertheless, there is still no consensus on the etiology of CM.^[4]

The craniovertebral junction is a site where many structural anomalies can be observed, and the need for surgery is frequent. The risk of vertebral artery (VA) injury during craniovertebral junction surgery for various indications has been reported as 4.1% in different series.^[5] Damage to the VA during surgery in this region is rare; however, when it occurs, it causes serious complications. The most significant factors causing injury are the variable anatomical location of the VA groove (VAG) or the proximity of the surgical procedure to the VA.^[6] Therefore, a detailed evaluation of the VAG with CT before surgery would significantly reduce the risk of perioperative complications.^[7]



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Considering VA-related injuries occurring during surgical intervention, it has been shown that two anatomical variations are the most common causes. One is the presence of a high-riding vertebral artery (HRVA), and the other is the presence of a narrow C2 pedicle. The prevalence of HRVA has been reported as 14.5–18% in different series, and the presence of a narrow C2 pedicle has been reported as 9.5–32% on average.^[8]

In the literature, HRVA has been defined as a C2 isthmus height of less than 5 mm and/or an internal height of less than 2 mm measured 3 mm lateral to the border of the spinal canal. Internal height is the shortest distance from the outer margin of the C2 transverse foramen to the surface of the superior articular process, and isthmus height is the thinnest part of the C2 isthmus (Fig. 1).^[9]

The aim of this study is to analyze the patients operated on for Chiari malformation type 1 in our clinic for the presence of a high-riding vertebral artery and to investigate the effect of this parameter on surgery.

MATERIALS and METHODS

This study was approved by the Medical Specialty Education Board of our hospital (TUEK) on 30.12.2020 with the number E-62977267-903.99. This study was conducted in accordance with the principles of the Helsinki Declaration. All of the patients underwent surgery for Chiari malformation type 1 between 2018 and 2020 in our clinic. The cases were retrospectively evaluated using patient files and preoperative CT imaging of the cervical spine.

The study included 15 patients, of whom 4 were female and 11 were male. Cervical spine CT scans, routinely performed for preoperative surgical planning, were analyzed bilaterally in all cases.

Routine sagittal cervical CT images obtained in 3-millimeter thin sections were used for computed tomography measurements. C2 isthmus height and internal height of the lateral mass were measured bilaterally for each case (Fig. 1).

IBM SPSS Statistics 22 (IBM SPSS, Turkey) was used for statistical analyses in the evaluation of the findings obtained in the study. The McNemar test was used to compare qualitative data. The significance level was assessed at $p < 0.05$.

RESULTS

In this study, computed tomography images of 15 patients who were operated on for CM-1 in our clinic were analyzed bilaterally. The mean age was 38.9 years (range 17–63). Of

the cases, 4 were male and 11 were female. No exclusion criteria were used since the cases were evaluated using radiologic images. Based on CT measurements, the mean isthmus height was 4.76 mm (range, 1.1–9.31 mm; standard deviation [SD], 1.31), and the mean internal height was 4.15 mm (range, 1.1–9.31 mm; standard deviation [SD], 1.31) (Table 1).

One (3.3%) of 30 C2 lateral mass internal height measurements indicated a high-riding vertebral artery on the right side, whereas a high-riding vertebral artery was found in 13 of 30 measurements according to C2 isthmus height measurements (43.3%). Of these, 3 were right-sided, 2 were left-sided, and 4 were bilateral (Table 2). In other words, 9 out of 15 patients displayed HRVA based on C2 isthmus height measurements.

The incidence of a high-riding vertebral artery was 46.7% on the right side and 40% on the left side. There was no statistically significant difference in the presence of a high-riding vertebral artery between the right and left sides based on the isthmus measurements ($p > 0.05$) (Table 3) (Fig. 2).

During the surgical intervention, there were no complications related to HRVA in any of the patients.



Figure 1. a: C2 vertebra internal height (the shortest distance from the outer margin of the C2 transverse foramen to the surface of the superior articular process); b: C2 vertebra isthmus height (the thinnest part of the C2 isthmus)

Table 1. Distribution of demographic and clinical characteristics of the cases

	Min-max	Median	Mean±SD	n	%
Age	17.0–63.0	41.0	38.9±13.6		
Gender					
Female				11	73.3
Male				4	26.7
Side					
Right				15	50.0
Left				15	50.0
Internal height					
Right	1.90–6.60	4.10	4.06±1.11		
Left	2.60–5.30	4.20	4.25±0.79		
Total	1.90–6.60	4.20	4.16±0.95		
Isthmus height					
Right	3.00–6.90	5.50	5.07±1.13		
Left	3.60–7.10	5.20	5.19±0.97		
Total	3.00–7.10	5.20	5.13±1.04		

SD: Standart deviation

DISCUSSION

Chiari malformation is a structural deformity first described by the Austrian pathologist Hans Chiari as herniation of the cerebellar tonsils into the foramen magnum. The most common type is CM-1. Herniation of the cerebellar tonsils more

Table 2. Internal height and isthmus height measurements of 15 cases from the right and left sides

Case no	C2 internal height R	C2 isthmus height L	C2 internal height R	C2 isthmus height L	HRVA (+) side
1	3.7	4.2	5.3	5.5	Right
2	3.2	4.0	3.9	4.4	Bilateral
3	3.5	6.2	3.3	6.0	
4	3.1	5.5	3.4	5.2	
5	4.0	4.4	4.2	5.2	Right
6	1.9	3.0	2.6	4.7	Bilateral
7	4.7	5.8	5.1	6.0	
8	5.2	5.6	4.7	4.3	Left
9	4.3	5.8	4.2	5.2	
10	4.5	4.8	4.1	4.2	Bilateral
11	4.6	6.9	4.8	7.1	
12	2.9	3.7	4.6	3.6	Bilateral
13	4.6	6.1	3.5	5.1	
14	6.6	6.1	5.0	4.5	Left
15	4.1	4.0	5.1	6.8	Right

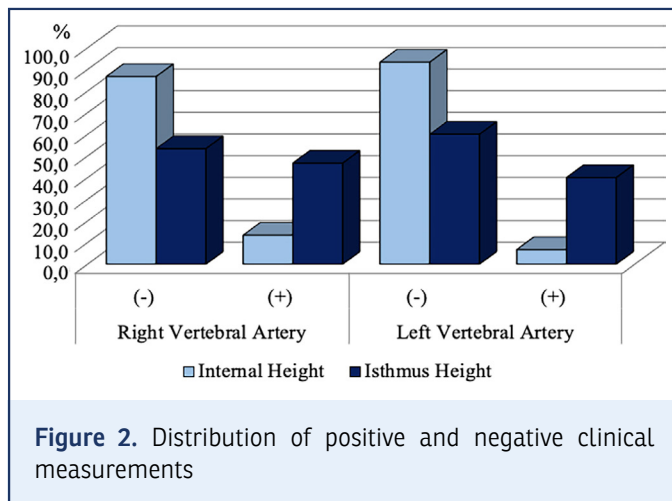
HRVA: High-riding vertebral artery

than 5 mm into the spinal canal is defined as CM-1.^[10] Different studies have reported incidence rates ranging between 0.56% and 1%. The pathology is usually asymptomatic and rarely requires surgical intervention.^[11,12]

Table 3. Distribution of comparative clinical measurements of the cases

	Internal height				Isthmus height				p
	Mean±SD	n	%	Median	Mean±SD	n	%	Median	
Total	4.16±0.95			4.20	5.13±1.04			5.20	0.000 ^w
Right	4.06±1.11			4.10	5.07±1.13			5.50	0.002 ^w
Left	4.25±0.79			4.20	5.19±0.97			5.20	0.011 ^w
Right/left difference p			0.363 ^w				0.363 ^w		
Total HRVA									
(-)		27	90.0			17	56.7		0.002 ^N
(+)		3	10.0			13	43.3		
Right HRVA									
(-)		13	86.7			8	53.3		
(+)		2	13.3			7	46.7		
Left HRVA									
(-)		14	93.3			9	60.0		
(+)		1	6.7			6	40.0		

^w: Wilcoxon test; ^N: Mc Nemar test. SD: Standart deviation; HRVA: High-riding vertebral artery



CM-1 may negatively affect a person's quality of life and cause symptomatic problems that might require surgical intervention. Surgical treatment is advised, especially in cases with signs of brainstem involvement, dysesthesia, and respiratory distress, accompanied by syringomyelia. The standard surgical procedure for Chiari malformation is posterior fossa decompression, C1 laminectomy, and partial C2 laminectomy in cases where the cerebellar tonsils descend to the level of the C2 vertebra. Additional interventions, such as opening or not opening the dura and occipitocervical fusion, can be included in the surgical technique.^[13] However, there is still no consensus on the effect of different surgical techniques on postoperative status.

Excessive decompression of the foramen magnum with suboccipital craniectomy may cause cerebellar ptosis, and inadequate decompression may prevent clinical improvement.^[14] Despite different opinions regarding the size of the craniectomy to be performed, Batzdorf et al.^[15] recommended decompression of the craniectomy with a height of 15–20 mm and a width of 20–25 mm, leaving bone to support the midline cerebellar hemispheres, excision of the C1 vertebra 10 mm lateral to the midline, and, if necessary, excision of the superior C2 lamina, taking care to preserve the spinous processes and ligaments. Measurement of the distance between two vertebral arteries above the C1 lamina is a key step in Chiari type 1 surgery. However, to our knowledge, the existence of HRVA in Chiari type 1 patients has not been reported in the literature previously.

In recent years, the increasing number of surgeries performed in the cranio-cervical region has increased the significance of the anatomy and variations of the vertebral artery.

In all surgical procedures involving the craniocervical junction, such as Chiari malformation, it is essential to understand the anatomy of the C1 and C2 vertebrae, referred to as "atypical vertebrae," and their relationship with the vertebral artery for surgical safety. The vertebral artery typically has four segments, three of which are extracranial and one intracranial. It leaves the subclavian artery, passes anterior to the C7 transverse process, and ascends superomedially within the transverse foramen of the C6 vertebra (V1 segment). The V2 segment extends vertically in the transverse foramina up to the level of the C2 vertebra. It then turns laterally and enters the transverse foramen of the C1 vertebra. After leaving the transverse foramen of the atlas, it continues posteriorly and extends horizontally superior to the posterior arch of the atlas (V3 segment). Then it pierces the dura posterior to the atlantooccipital membrane and continues into the intracranial space with the V4 segment through the foramen magnum. Complications associated with VA injury, which may have fatal consequences, have been reported in the literature with a frequency of 4.1%–8.2%, especially in surgeries where stabilization of the craniovertebral junction is added.^[16]

Turgut et al.^[17] analyzed 194 cases reported in 72 studies conducted between 1962 and 2021 and reported laceration of the artery (41.24%), pseudoaneurysm (16.49%), and VA dissection (5.67%) .

Intraoperative unilateral injury of the vertebral artery may remain asymptomatic, or it may lead to hemorrhage, infarction of the brainstem, and unpredictable neurologic deficits depending on the compensation capacity from the contralateral vertebral artery.

Accompanying vascular anomalies are not common in patients with CM-1. However, in their cadaveric study, Boggio et al.^[18] reported a case in which CM-1 was accompanied by a bi-hemispheric PICA variation, an extremely rare pathology. They warn to be cautious of unilateral PICA injury during posterior fossa decompression.

Regarding VA-related injuries that occur in surgical interventions for the craniocervical junction, two anatomical variations have been reported to be the most common causes. One is the presence of a high-riding vertebral artery, and the other is the presence of a narrow C2 pedicle. The definition and prevalence of HRVA are well-defined in the literature. HRVA has been defined as a C2 isthmus height (thinnest part of the C2 isthmus) of less than 5 mm and/or an internal height (shortest distance from the outer margin of the C2 transverse foramen to the surface of the

superior articular process) of less than 2 mm measured 3 mm lateral to the border of the spinal canal.

In addition, Cine et al.^[19] in their study reporting 8 patients, mentioned that the C2-3 transfacet screw procedure is an extremely effective and reliable technique as an alternative to other techniques and salvage methods, especially in selected patients with high-riding vertebral artery and a thin, narrow C2 pedicle. In our study, no case required craniospinal fusion.

In a meta-analysis study conducted by Klepinowski et al.^[9] with 3,126 patients, the prevalence of HRVA was found to be 25.3%. In patients with a diagnosis of rheumatoid arthritis, it was significantly higher than in those without rheumatoid arthritis. No significant difference was found regarding genders and geographical regions. No significant right-to-left dominance was found, whereas unilateral and bilateral HRVA were found in 70% and 29.7% of the cases, respectively. There was no patient diagnosed with RA in our cases. In the measurements of 15 patients, 9 cases (60%) were found to have HRVA. Yamazaki et al.^[20] reported 5 out of 31 cases (16.1%) of unilateral HRVA in their study. Unlike the literature, our results revealed that only one patient had HRVA based on the C2 internal height measurement, whereas 9 patients were identified with HRVA based on isthmus height measurements. Since HRVA can be defined based on either of these measurements, we consider that this difference may be related to the small number of cases.

Consistent with the literature, no significant right-left or laterality distinction was found for the presence of HRVA in our series. In our patients, there were three right-sided, two left-sided, and four bilateral HRVAs, and there was no statistically significant difference between the right and left sides.

We observed that the majority of studies in the literature were related to posterior craniocervical instrumentation surgery and the presence of HRVA. Although the C2 vertebra is not involved in the standard surgical procedure for CM-1 patients, it is important to avoid C1 laminectomy. Surgeons should be aware of vascular variations, especially when operating in the far lateral regions of the craniovertebral junction. In complicated CM-1 patients, more complex surgical procedures might be needed, such as C1-C2 fixation. Especially in these kinds of patients, investigating vascular variations and anatomical considerations is more important in preoperative planning.

In specific cases of CM-1, considering the frequency of HRVA is crucial in preventing catastrophic vascular complications during surgery. Preoperative imaging tech-

niques, such as vertebral CT angiography, and intraoperative neuromonitoring in such patients would help prevent surgical complications.

We acknowledge that the single-center, retrospective nature of our study and the limited number of cases are its main limitations. Our findings indicate that the results of our study are consistent with measurements from larger series evaluating HRVA using similar methodologies. The frequency of HRVA in patients with CM-1 does not differ from that of the general population. However, we emphasize that in cases requiring cranio-cervical fusion, this anatomical variation should be considered to minimize the risk of surgical complications. These findings highlight the need for multi-center, prospective studies with larger case series to provide more comprehensive insight into the significance of HRVA, especially in complex CM-1 cases.

CONCLUSION

This study aimed to investigate the frequency of high-riding vertebral artery and its effect on surgical planning in patients who underwent decompression of the posterior fossa due to CM-1. Despite the relatively high incidence of HRVA in our case group, none of the patients experienced any complications related to VA injury. In conclusion, we believe that investigating the presence of HRVA in preoperative planning in CM-1 patients should be considered to avoid possible complications.

Disclosures

Ethics Committee Approval: The study was approved by the Haydarpaşa Numune Training and Research Hospital Medical Specialty Education Board of our hospital (TUEK) Ethics Committee (No: E-62977267-903.99, Date: 30/12/2020).

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