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Obex position is associated with syringomyelia and use of posterior fossa decompression among patients with Chiari I malformation

Gabe Haller, PhD^{1,2}, Brooke Sadler, PhD², Timothy Kuensting, BA², Nivan Lakshman¹, Jacob K. Greenberg, MD,MSCI¹, Jennifer M. Strahle, MD¹, Tae Sung Park, MD¹, Matthew B. Dobbs, MD^{3,4}, Christina A. Gurnett, MD,PhD^{2,3,5}, David D. Limbrick Jr., MD,PhD¹

¹Department of Neurological Surgery, Washington University School of Medicine;

²Department of Neurology, Washington University School of Medicine;

³Department of Orthopaedic Surgery, Washington University School of Medicine;

⁴Shriners Hospital for Children, St. Louis, Missouri

⁵Department of Pediatrics, Washington University School of Medicine;

Abstract

OBJECTIVE—Chiari I malformation (CM-I) has traditionally been defined by measuring the position of the cerebellar tonsils relative to the foramen magnum. The relationships of tonsillar position to clinical presentation, syringomyelia, scoliosis, and the use of posterior fossa decompression (PFD) surgery have been studied extensively and yielded inconsistent results. Obex position has been proposed as a useful adjunctive descriptor for CM-I and may be associated with clinical disease severity.

METHODS—A retrospective chart review was performed of 442 CM-I patients with MRI who presented for clinical evaluation between 2003 and 2018. Clinical and radiological variables were measured for all patients, including presence/location of headaches, Chiari Severity Index (CSI) grade, tonsil position, obex position, clival canal angle, pB-C2 distance, occipitalization of the atlas, basilar invagination, syringomyelia, syrinx diameter, scoliosis, and use of PFD. Radiological measurements were then used to predict clinical characteristics using regression and survival analyses, with performing PFD, the presence of a syrinx, and scoliosis as outcome variables.

RESULTS—Among the radiological measurements, tonsil position, obex position, and syringomyelia were each independently associated with use of PFD. Together, obex position, tonsil position, and syringomyelia (area under the curve [AUC] 89%) or obex position and tonsil position (AUC 85.4%) were more strongly associated with use of PFD than tonsil position alone (AUC 76%) ($P_{\text{diff}} = 3.4 \times 10^{-6}$ and 6×10^{-4} , respectively) but were only slightly more associated

Correspondence: Gabe Haller: Washington University School of Medicine, St. Louis, MO. ghaller@wustl.edu.

Author Contributions

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than obex position alone (AUC 82%) ($P_{\text{diff}} = 0.01$ and 0.18 , respectively). Additionally, obex position was significantly associated with occipital headaches, CSI grade, syringomyelia, and scoliosis, independent of tonsil position. Tonsil position was associated with each of these traits when analyzed alone but did not remain significantly associated with use of PFD when included in multivariate analyses with obex position.

CONCLUSIONS—Compared with tonsil position alone, obex position is more strongly associated with symptomatic CM-I, as measured by presence of a syrinx, scoliosis, or use of PFD surgery. These results support the role of obex position as a useful radiological measurement to inform the evaluation and potentially the management of CM-I.

Keywords

obex position; Chiari I malformation; scoliosis; syringomyelia; headaches; tonsil position; posterior fossa decompression surgery

CHIARI I malformation (CM-I) has traditionally been defined by the presence of greater than 5 mm of tonsillar ectopia below the foramen magnum, though this definition is based primarily on historical precedent rather than clinical evidence.¹ Questioning the utility of this definition, the relationship between tonsil position and measures of disease severity, such as syringomyelia and surgical treatment with posterior fossa decompression (PFD), has been inconsistently reported in the CM-I literature.^{2–15} These results emphasize the need for improved understanding of the association between radiological craniovertebral junction (CVJ) parameters and clinical disease in cases of CM-I.

Various terms have been used to describe a spectrum of patients with a complex CM-I phenotype who have concurrent CVJ or posterior fossa pathology and/or variant anatomy.¹⁶ Obex position is one CVJ measure that has been investigated in CM-I, and an obex position below the level of the foramen magnum, in combination with tonsillar ectopia, has been classified by some authors as a further disease subset, Chiari 1.5 (CM-1.5).^{4,16} However, an obex position is inconsistently reported in the CM-I literature, and the degree to which CM-1.5 represents a distinct disease subset is unclear.^{4,17} While limited evidence in adult CM-I patients suggests obex position may predict symptom resolution after PFD,¹⁵ the clinical importance of obex position in children remains to be elucidated.

Given this gap in the existing literature, the objective of this study was to investigate the association of obex position, tonsillar position, and other CVJ morphometric measurements with measures of clinical disease severity in children with CM-I.

Methods

Clinical and Radiological Measurements

After IRB approval from Washington University in St. Louis, a retrospective chart review was performed on all pediatric patients who presented to the outpatient clinic and were diagnosed with CM-I at St. Louis Children's Hospital between 2003 and 2018. Patients were diagnosed based on 5 mm or more of tonsillar ectopia demonstrated on imaging. Patients diagnosed with Chiari II malformations were excluded. Three patients with reported

tethered cord were included. A total of 599 patients with a tonsil position of at least 5 mm below the level of the foramen magnum were identified, of whom 442 had a high enough quality preoperative brain MR image available for review. Most patients had MR images that included the spine. If multiple MR images were done, the oldest study was always used to ensure presurgical symptom characterization and for consistency. Manual review of MR images was performed to identify tonsil position¹² (negative values below the foramen magnum), obex position (the distance of the obex from the foramen magnum, as defined by the basion-opisthion line with negative values below the foramen magnum),¹⁶ syrinx diameter (maximum anteroposterior distance or lateral distance of the syrinx cavity; no size cutoff was used), basilar invagination,¹⁸ occipitalization of the atlas (defined as fusion of C1 to the base of the skull), pB-C2 distance,^{6,19,20} and clival canal angle.¹⁸ Syringomyelia was analyzed as a baseline radiological independent variable and also as an outcome measure, or dependent variable.

Charts were reviewed to determine the presence, severity, and type of headaches; clinically diagnosed scoliosis; age at CM-I diagnosis; age at PFD (if applicable); age at initial MRI; height; weight; family history; and Chiari Severity Index (CSI) grade.⁷ The CSI is an integrated clinical and radiological severity index that has been used to classify disease severity in children with CM-I⁷ and includes aspects of headache symptoms, bulbar symptoms related to brainstem compression, and spinal cord pathology. Our primary outcome measurements were PFD surgery and syringomyelia. Secondary outcomes analyzed were scoliosis, CSI grade, headaches (presence or absence, as noted in patients' charts), and primary headache location (occipital, frontotemporal, or unspecified).

All imaging studies and medical records were reviewed by a single trained reviewer. Imaging controls were identified among patients seen for headaches at pediatric neurology clinics at St. Louis Children's Hospital who were not found to have tonsillar herniation or other intracranial pathology.

Statistical Analysis

Univariate and multivariate analyses was performed with logistic regression using the GLM function in R. The correlation between continuous variables was determined by linear regression using the GLM function in R. Receiver operating characteristic (ROC) curves were obtained for the ability of each variable to predict surgery using the pROC package in R and compared pairwise using De-Long's test for two ROC curves. Multiple variables were combined using logistic regression to obtain fitted values for use in ROC estimates. Kaplan-Meier survival curves were created using the OIsurv package in R.

Results

Of the 442 pediatric CM-I patients who were included in this study, the mean age was 13 ± 6 years at presentation and 240 of 442 (54%) were female. Of the 442 patients, 113 (27%) had syringomyelia at presentation and 137 (31%) ultimately underwent PFD. Among those with syringomyelia, the mean syrinx size was 5.85 ± 3.8 mm. All patients had tonsil positions greater than 5 mm below the foramen magnum. The mean tonsil position was -9.4 ± 5.2 mm (range -5 to -28 mm) below the foramen magnum. The mean obex position among CM-I

patients was 2.3 ± 5.8 mm above the foramen magnum. Scoliosis was diagnosed in 15% of CM-I patients. We tested associations with all variables and measurements mentioned in *Methods* and followed up further with significant results. Indications for performing PFD were assessed by an attending physician (D.D.L.). Among patients undergoing PFD, 43% had a primary indication for surgery of syringomyelia, 40% had a primary indication for surgery of headaches, 4% had a primary indication for surgery of scoliosis, and the remaining 13% of patients had various primary indications for surgery, including vision changes, peripheral weakness, dysphasia, neck pain, and sleep apnea, among others. No patients were offered PFD based on tonsil position alone, and no patients in this study had an obex position measured prior to surgery.

Predicting PFD Using Clinical and Radiological Characteristics

The association between the analyzed clinical and radiological variables and treatment with PFD is shown in Table 1. In the univariate analysis, tonsil position, obex position, syringomyelia, CSI grade, and occipital headaches were associated with use of PFD (p value range = 4×10^{-4} to 1.6×10^{-27}) (Table 1).

Obex and Tonsil Position—Since tonsil position is a well-established measurement of CM-I and is associated with use of PFD, we determined if the addition of obex position would improve the association of PFD surgery use. Obex position and tonsil position were correlated in this data set ($r^2 = 0.32$, $p = 5 \times 10^{-35}$) (Fig. 1A), with the majority of surgical cases having both moderate-severe tonsil position (< -10 mm, i.e., 10 mm below the foramen magnum) and obex position (< 0 mm, i.e., below the foramen magnum)—in other words, they had CM-1.5. When these significant measures were combined in a single regression analysis, tonsil position, obex position, and syringomyelia remained independently associated with PFD. Together, these factors combined were associated with the use of PFD with an area under the curve (AUC) of 89%. This was significantly better than when tonsil position (AUC 76%) or obex position (AUC 82%) was used alone ($P_{\text{diff}} = 3.4 \times 10^{-6}$ and $P_{\text{diff}} = 0.01$, respectively) (Fig. 1B). This was also evident in that obex position was significantly lower among patients undergoing decompression (Fig. 1B) ($p = 7.7 \times 10^{-25}$) and that obex position and tonsil position together were more significantly associated with the use of PFD than tonsil position alone (AUC of 85.4% vs 76%; $P_{\text{diff}} = 6 \times 10^{-4}$) (Fig. 1C).

To determine if obex position had a significant and independent association with use of PFD, we stratified patients by both obex position and tonsil position after dichotomizing for each within the data set. Individuals were categorized as either having a low obex position (< 0 mm), moderate-severe tonsil ectopia (< -10 mm), neither, or both. Survival analysis with PFD as the outcome variable revealed that patients with moderate-severe tonsil position (< -10 mm) and normal obex position (> 0 mm) and patients with a low obex position (< 0 mm) but mild tonsil position (> -10 mm) were more likely to have undergone PFD ($P_{\text{diff}} = 2 \times 10^{-6}$ and 5×10^{-4} , respectively) than patients with a normal obex position and mild tonsil position. Similarly, patients with both a low obex position and a moderate-severe tonsil position were more likely to have undergone PFD compared to patients with a mild tonsil position and a normal obex position ($P_{\text{diff}} = 2 \times 10^{-14}$) and compared to patients with

either a low obex position but a mild tonsil position or a moderate-severe tonsil position but a normal obex position (> 0 mm) ($P_{\text{diff}} = 0.01$) (Fig. 1D).

Syringomyelia—Since syringomyelia is a major factor in surgical decision-making for PFD, we investigated the relationships among syringomyelia, tonsil position, and obex position. We found that, after grouping patients by the presence of a syrinx, both obex position and tonsil position remained significantly associated with use of PFD among patients without syringomyelia ($p = 3.5 \times 10^{-6}$ and 2.9×10^{-8} , respectively [logistic regression]) but found no relationship between either obex position ($p = 0.11$, logistic regression) or tonsil position ($p = 0.07$, logistic regression) among patients with syringomyelia, as nearly all patients with syringomyelia underwent PFD in this data set. We performed this same analysis using syringomyelia as the outcome and found that the presence of a syrinx is predicted best by both obex position and tonsil position (AUC 71%; $p = 6.2 \times 10^{-3}$ compared to tonsil alone), but obex position alone still predicted the presence of a syrinx with a higher AUC than herniation alone (AUC 63% for obex position vs 56% for tonsil position) (Fig. 1E).

Predicting Syringomyelia Using Clinical and Radiological Characteristics

We next evaluated clinical or radiological measurements associated with the presence of a syrinx. Of the measured characteristics, tonsil position and obex position were the only factors that were significantly associated with syringomyelia. CSI grade was not tested as it is partially defined by the presence of syringomyelia. Including both tonsil position and obex position in a single regression analysis revealed that only obex position was significantly associated with syringomyelia independent of tonsil position, with an AUC of 70.2%, a sensitivity of 70%, and a specificity of 70%. We next stratified patients into groups based on the severity of tonsil and obex positions. There were similar rates of syringomyelia in individuals with mild obex position (≤ 0 mm) and moderate-severe tonsil position (≤ -10 mm) compared to those with mild obex position (> 0 mm) and mild tonsil position (> -10 mm). Similarly, individuals with a low obex position (< 0 mm) and moderate to severe tonsil position (≤ -10 mm) had similar rates of syringomyelia to patients with a severe obex position and a mild tonsil position (Fig. 2). Basilar invagination, occipitalization of the atlas, pB-C2 distance, and clival canal angle were not associated with any of the primary outcomes and were not correlated with tonsil position or obex position.

Relationship Among Obex Position, Scoliosis, and Syringomyelia

As scoliosis is an important comorbidity in CM-I patients with syringomyelia, we investigated the relationship among obex position, scoliosis, and syringomyelia in this data set. First, we replicated previous findings that scoliosis is significantly more common in CM-I patients with syringomyelia compared to CM-I patients without syringomyelia ($p = 4.74 \times 10^{-9}$). Next, we found that obex position was significantly lower in CM-I patients with scoliosis compared to those without (-1.2 mm vs 2.7 mm; $p = 2 \times 10^{-5}$). Tonsil position was not different between CM-I patients with or without scoliosis (-9.8 mm vs -10.6 mm; $p = 0.27$). When we stratified patients by scoliosis and obex position, we found an increased rate of syringomyelia among patients with a low obex position (< 0 mm) but no scoliosis (43% of patients), scoliosis but a normal obex position (59% of patients),

or both a severe obex position and scoliosis (80% of patients) compared to patients with neither scoliosis nor a severe obex position (13% of patients) (Fig. 3). In order to determine if obex position was related to syringomyelia risk through its relationship to scoliosis, we stratified patients by obex position and syringomyelia and looked at the rate of scoliosis in these subsets of patients. We found no relationship between obex position and scoliosis when stratified by syringomyelia, or to put it another way, the obex is only associated with scoliosis through its association with syringomyelia.

Impact of Tonsil and Obex Positions on CM-I–Related Symptoms

To better understand the relationship between radiological characteristics and symptoms of patients with CM-I, we next determined if any of these measurements were associated with CM-I–related symptoms including presence of headaches, headache location, and CSI score.⁷ Obex position, but not tonsil position, was correlated with the presence of occipital headaches ($p = 8 \times 10^{-4}$ [logistic regression]) but not the presence of any kind of headache ($p > 0.05$), with a more severe obex position occurring more often in patients with occipital headaches (Fig. 4 left). Similarly, in a multivariate analysis including both obex position and tonsil position, patients with more severe obex positions were less likely to have CSI grade 2 (small or no syrinx and only frontotemporal or no headaches) ($p = 2 \times 10^{-3}$ [logistic regression]) (Fig. 4 right). This is consistent with our finding that a combination of obex position and the presence of a syrinx are the most strongly associated factors with CM-I severity as measured by use of PFD.

Discussion

We evaluated a large cohort of CM-I patients to determine both clinical and radiological variables associated with CM-I pathology, including syringomyelia, clinical symptoms, and use of PFD. We show that 3 radiological variables—tonsil position, obex position, and syringomyelia—were independently associated with use of PFD surgery in CM-I. However, importantly, obex position was associated with both syringomyelia and scoliosis, independent of tonsil position. Although tonsil position and obex position were correlated in our group of CM-I patients, obex position was independently associated with syringomyelia, scoliosis, occipital headaches, and CSI grade and may represent an important adjunct in the clinical decision-making process for CM-I patients at presentation, since many neurosurgeons do not currently independently consider obex position when determining which patients require PFD. However, we believe that surgeons are unknowingly selecting those patients with the worst obex positions for surgery, as all variables surgeons currently consider are associated with PFD through obex position. For example, our finding that poor obex position is independently associated with occipital headaches would increase the frequency of those with poor obex positions receiving PFD. The converse then is true as well—that those patients with less severe obex positions are the ones more often not offered surgery.

Though controversial, many clinicians use a threshold of less than -5 mm for cerebellar tonsillar position of one tonsil to diagnose CM-I.²¹ This cutoff is based on measurements of average tonsil position in control and CM-I populations.^{2,3} Furthermore, several variables

have been associated with clinical symptoms. For example, the presence and size of a syrinx increases the risk of scoliosis,^{5,6} and syringomyelia is more common in patients with a greater degree of tonsillar descent.¹³ However, the relationship between tonsil position and clinical disease has been inconsistently reported.^{2,3,11}

Tonsil position has a notoriously inconsistent relationship with clinical outcome, evidenced by the fact that up to 3.4% of adults undergoing MRI would qualify for a CM-I diagnosis based on tonsil position alone; however, only 0.01%–0.04% of these patients would be symptomatic.²² Recognizing the heterogeneity among CM-I patients, CM-1.5 was defined as the presence of an obex position below the foramen magnum.^{4,16} However, this diagnosis is applied inconsistently, and many patients with a low obex position may not be grouped into this category. Furthermore, unlike tonsil position, there are no data on the normal distribution of the obex, and its clinical impact remains poorly defined.¹¹ Thus, it is important to evaluate this matter in a large cohort of CM patients. As CM-I is likely a more heterogeneous entity than appreciated, there is a need to evaluate other variables as well. For example, the presence of gait imbalance and motor deficits was recently found to predict worse clinical and radiological outcomes after surgery in an adult population,¹⁵ and increased tonsillar ectopia has been shown to be associated with a more posterior odontoid angulation²³ and displaced obex.¹⁰ Using multiple predictive variables, we are the first to report, with statistical confidence, that the position of the obex is more strongly associated with CM-I symptoms than tonsil position, as well as the first to describe in detail the relationship among obex position, the presence of a syrinx, and clinical disease. While it may seem logical that patients with a low obex position have greater clinical symptom severity, we observed a low obex position in 32% of our sample, suggesting that the true prevalence of CM-1.5 is much higher than previously thought. This figure jumps to 63% of patients with a low obex among patients undergoing PFD.

Though previous studies have shown that the presence of a syrinx is independently associated with scoliosis,¹⁴ this is the first study to report the association between obex position and scoliosis. Our data suggest, however, that this association is driven through the association of both obex and scoliosis with syringomyelia. We also found an association between occipital headaches and low obex position. The majority of CM-I patients in our cohort reported experiencing frequent headaches. Similarly, CSI grade contains components of headache symptomatology and syringomyelia, both factors correlated with obex position. Given the associations of low obex position with syringomyelia and scoliosis, obex position may factor into the decision to obtain an MR image of the spine as well. Building on the existing literature,^{2,3, 6, 11, 16, 17, 19,24} these results suggest that obex position may be a key predictor of clinical disease in CM-I patients and can serve as an important element for evaluating and classifying this heterogeneous patient population, and the results also indicate that obex position is more strongly associated with CM-I–related symptoms than tonsil position.

Limitations

There are several limitations to this study. First, this study is retrospective, which may have biased the collection and evaluation of certain data elements. Second, all MRI measurements

were performed at a single time point in any individual, and therefore our analysis did not evaluate variability in obex position over time or how well patients did postoperatively. Third, our primary outcome, undergoing PFD surgery, was based on individual surgeon judgment, which may have biased this measure of clinical disease severity. Fourth, many of the measurements performed in this study can be challenging given the quality of the MR images being screened, and measurement of several features was not possible after PFD surgery, given the alterations to the structures involved. Finally, we did not assess the relationship between obex position and postoperative change in CM-I–related pathology, such as clinical symptoms, syringomyelia, and scoliosis. In future work, we plan to evaluate the relationship between obex position and these key postoperative outcomes.

Conclusions

In this study we defined the relationships among obex position, tonsil position, and syringomyelia and CM-I symptoms, use of surgery, and scoliosis. Compared to tonsil position alone, obex position is more strongly associated with CM-I severity, as measured by the use of PFD and presence of a syrinx and scoliosis. While further validation is needed, these results suggest that obex position may be a useful radiological measurement in the management of CM-I and that CM-1.5 needs to be a more widely adopted and diagnosed category of Chiari malformation.

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Disclosures

Dr. Limbrick reports receiving support of non-study-related clinical or research efforts that he oversees from Medtronic, Inc., and Microbot Medical, Inc.

ABBREVIATIONS

AUC	area under the curve
CM-I	Chiari I malformation
CSI	Chiari Severity Index
CVJ	craniovertebral junction
PFD	posterior fossa decompression
ROC	receiver operating characteristic

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FIG. 1.

Obex position improves surgical risk prediction among CM-I patients. **A:** Correlation between obex position and tonsil position among CM-I patients dichotomized by surgical treatment. **B:** Histogram of obex position among CM-I patients with and without decompression surgery. **C:** ROC curves showing PFD surgery is predicted best by both obex position and tonsil position. **D:** Kaplan-Meier curve comparing the proportion of CM-I patients receiving PFD surgery among individuals dichotomized by obex position (≥ 0 mm), tonsil position (≥ -10 mm), or both obex position and tonsil position. **E:** ROC curves showing syringomyelia is predicted best by both obex position and tonsil position.

FIG. 2.

Obex position is associated with syringomyelia independently of tonsil position among patients with CM-I. Relationship between dichotomized obex position and syringomyelia risk stratified by tonsil position. Mild obex and mild tonsil position (TP) (obex > 0 and TP > -10); mild obex and severe tonsil position (obex > 0 and TP ≤ -10); low obex and mild tonsil position (obex ≤ 0 and TP > -10); low obex and moderate-severe tonsil position (obex ≤ 0 and TP ≤ -10). *p < 0.01, ***p < 0.0001.

NS = not significant.

FIG. 3.

Obex position is associated with syringomyelia independently of scoliosis among patients with CM-I. Relationship between dichotomized obex position and syringomyelia risk stratified by either tonsil position or scoliosis. Mild obex and mild tonsil position (obex > 0 and TP > -10); mild obex and moderate-severe tonsil position (obex > 0 and TD < -10); low obex and mild tonsil position (obex ≤ 0 and TD > -10); low obex and moderate-severe tonsil position (obex ≤ 0 and TD < -10). *p < 0.01, ***p < 0.0001.

TABLE 1.

Effects of radiological and clinical characteristics on risk of PFD

Trait	All CM-I	Surgery (n = 137)	No Surgery (n = 294)	OR (95% CI)	p Value
Sex				0.97 (0.98–1.03)	0.36
Male	202 (46)	62 (45)	135 (46)		
Female	240 (54)	75 (55)	159 (54)		
Age (yrs)	13 ± 6	14 ± 6	12 ± 6	1.01 (0.99–1.03)	0.19
Surgery	137 (31)				
Tonsil position (mm)	−9.4 ± 5.2	−13.3 ± 5.6	−7.6 ± 4.0	1.3 (1.21–1.35)	1.2 × 10^{−23}
Obex position (mm)	2.3 ± 5.8	−1.5 ± 6.1	4.7 ± 4.4	1.23 (1.17–1.29)	7.7 × 10^{−25}
Syringomyelia	113 (27)	81 (60)	30 (11)	11.8 (6.99–19.88)	1.6 × 10^{−27}
Syrinx size (mm)	5.85 ± 3.8	6.52 ± 3.6	4.46 ± 2.8	1.3 (1.06–1.55)	0.008
Scoliosis	66 (15)	33 (24)	10 (3)	7.9 (3.71–16.73)	6 × 10^{−8}
CSI score					
1	91 (27)	51 (46)	39 (18)	3.77 (2.25–6.31)	4.4 × 10^{−7}
2	208 (62)	32 (29)	172 (79)	0.11 (0.06–0.19)	3.3 × 10^{−16}
3	34 (10)	28 (25)	6 (2)	11.6 (4.63–29.26)	1.8 × 10^{−7}
pB-C2 distance (mm)	6.5 ± 1.8	6.5 ± 1.9	6.6 ± 1.8	1.02 (0.91–1.14)	0.71
Basilar invagination	11 (2)	5 (4)	6 (2)	1.47 (0.40–5.33)	0.55
Headaches	217 (49)	81 (59)	131 (45)	1.56 (1.01–2.39)	0.04
Occipital headaches	88 (20)	44 (32)	43 (14)	2.44 (1.48–3.98)	3.9 × 10^{−4}
Occipitalization	30 (7)	14 (11)	16 (6)	1.88 (0.89–4.00)	0.10
Clival canal angle (°)	141 ± 12	139 ± 13	142 ± 11	0.99 (0.97–1.00)	0.15

Values are presented as the number (%) of patients or as the mean ± SD. Boldface type indicates statistical significance.