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ORIGINAL ARTICLE

MORPHOMETRIC ANALYSIS OF POSTERIOR FOSSA AND CRANIAL-VERTEBRAL JUNCTION IN PEDIATRIC CHIARI 1 MALFORMATION

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ABSTRACT

Introduction: Chiari malformations (CMs) are a group of disorders defined by anatomic anomalies of the cerebellum, brainstem, and cranial-vertebral junction (CVJ). We aimed to reveal the differences between pediatric CM-1 and control group according to volume, length and angle of cranium and CVJ.

Material and Methods: We retrospectively evaluated CM-1 patients who were admitted to our hospital between January 2012 and February 2016. Control group was choice from patients who did not have any intracranial pathology. We made necessary volume, length and angle evaluations with the help of special radiological programs.

Results: We evaluated 27 patients; 12 CM-1 cases and 15 control cases. Mean age in patient group was 12,66 ± 4,41 (2-17 years) years. Mean age in control group is 10,40 ± 4,62 (4-17 years) years. We found statistically significant in comparison of CD angle (p: 0,007) and C-SO angle (p: 0,017).

Conclusion: There is no difference between pediatric CM-1 patients and healthy group according to intracranial volumes, length and angle of cranial-vertebral junction. Clive-dental angle is narrower in pediatric CM-1 patients. This sharp passage leads to brain stem compression symptoms.

Keywords: Morphometry, posterior fossa, cranial-vertebral junction, Chiari malformation, type 1

Level of Evidence: Retrospective Clinical Study, Level III

INTRODUCTION

Chiari malformations (CMs) are a heterogeneous group of disorders that are defined by anatomic anomalies of the cerebellum, brainstem, and cranial-vertebral junction (CVJ), with downward displacement of the cerebellum, either alone or together with the lower medulla, into the spinal canal [28]. Chiari malformations were first described by John Cleland in 1883 [5,26]. Hans Chiari later classified them in 1891, into four groups. The pathology is a result of the underdevelopment of the para-axial mesoderm [22,24,30]. Consequently, the components of the posterior fossa outgrow the underdeveloped compartment and cause the herniation of the tonsils into the upper cervical spinal canal [33]. Several studies have attributed this insufficient posterior cranial fossa geometry to embryological defects in the paraxial mesoderm [20,22,34].

A fundamental knowledge of the normal anatomy of the cranial base, especially the foramen magnum and associated structures, is important to the clinician for accurate diagnosis and treatment of various diseases [16]. The cranial base has been noted for its ability to remain intact in cases where the rest of the cranium has been compromised and researchers have made use of that fact by analyzing sexually significant dimorphic trait for this anatomic region [12,14].

In this study, our purposes are 1) to establish whether the posterior fossa volume (PFV) is indeed different in individuals with pediatric CM-1than in healthy individuals, 2) to investigate the correlation within PFV, the area of foramen magnum (FMA) and intracerebral volume (ICV), 3) to understand the pathophysiology of CM-1.

MATERIAL AND METHODS

Patient population

We studied 12 patients and 15 control cases. A retrospective review of patients evaluated or operated for Chiari malformation type 1(CM-1) at the hospital between December 2012 and February 2016 was performed. The study group included patients all CM-1 with or without symptoms. We excluded the patients who do not have data to evaluate cranial volume and morphology. The control cases were included into study from patients admitted to hospital for headache or any reason without any intracranial pathology.

Definition of Chiari malformations

In classical classification [28-29];

- Chiari I malformation (CM-I) is characterized by abnormally shaped cerebellar tonsils that are displaced below the level of the foramen magnum,
- Chiari II malformation (CM-II) is characterized by downward displacement of the cerebellar vermis and tonsils, a brainstem malformation with beaked midbrain on neuroimaging, and a spinal myelomeningocele,
- Chiari III malformation (CM-III) is rare and combines a small posterior fossa with a high cervical or occipital encephalocele, usually with displacement of cerebellar structures into the encephalocele, and often with inferior displacement of the brainstem into the spinal canal,
- Chiari IV malformation (CM-IV) is now considered an obsolete term that describes cerebellar hypoplasia unrelated to the other Chiari malformations.

Radiological evaluation

The protocol for evaluating CMs included a lateral view radiograph of the CVJ. All patients underwent MRI (Sigma 1.5-Tesla; General Electric, Milwaukee, WI, USA). The T2-weighted MRI sequence was used for all measurements. Linear dimensions were derived using Extreme Pacs Workstation 1.5 software (Extreme PACS Healthcare, Ankara, Turkey). MRI of the CVJ at 5 mm intervals parallel to the orbitomental line was performed to determine a plane parallel to the foramen magnum (FM). The measurement of CD was performed on MRI sequences using the same software.

Measurement of volume

Calculation of spheroidal PFV was based on a simple spheroidal formula [13].

PFV=4/3 x ∏ x (X/2 x Y/2 x Z/2)

where: x is the anterior posterior measurement from the posterior clinoid process to the torcula; y is the height of the posterior fossa measured from the basion to the peak of the tentorium cerebelli; and z is the maximum width of the posterior fossa (Figure-1).

The ICV in children was calculated using a Dekaban spheroidal formula, which estimates the cranial volume in individuals up to20 years of age (t = thickness of the skull and scalp thickness) [1,18-19]:

ICV (cm^3) = 0,523 . (length – 2t) x (breadth – 2t) x (height – t) (Figure-2).

Measurement of FM area

The area of Foramen magnum was calculated using formula derived by Radinsky [27].

Radinsky's Formula (FMA) = 1/4 x FML x FMW

Where, (mathematical constant) = 22/7, FML = Foramen magnum length and FMW = Foramen magnum width (Figure-3).

Length of the clivus and sub-occiput

The length of the clivus (LoC) was defined as the distance from the topof the dorsum sellae to the basion, and the length of the subocciput (LoSO) was measured between the internal occipital protuberance and the opisthion [24] (Figure-4).

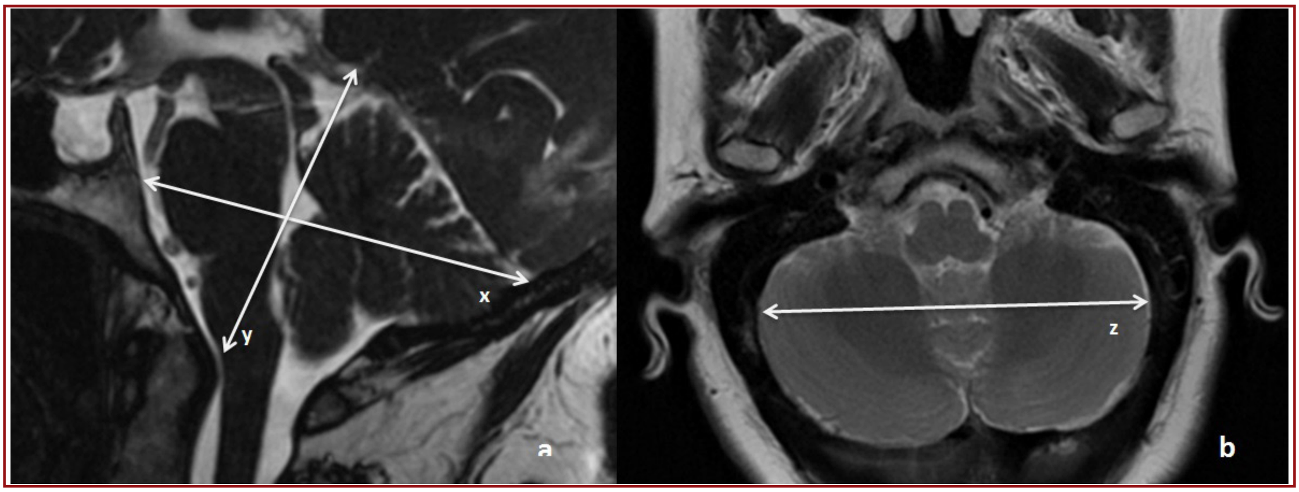

Figure-1. Spheroidal PFV is calculated using simple spheroidal formula.

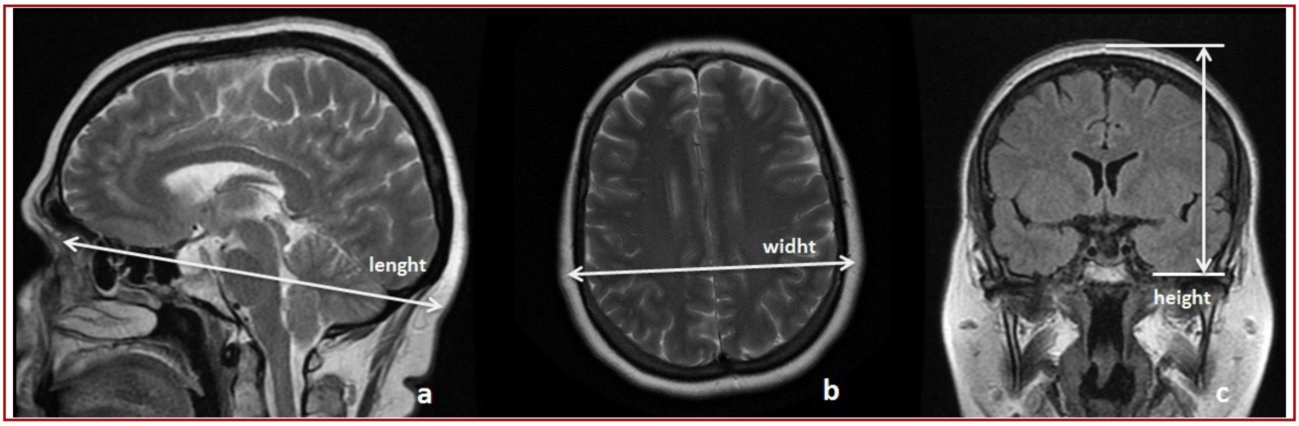

Figure-2. In children (<20 yrs), ISV was calculated using the Dekaban spheroidal formula

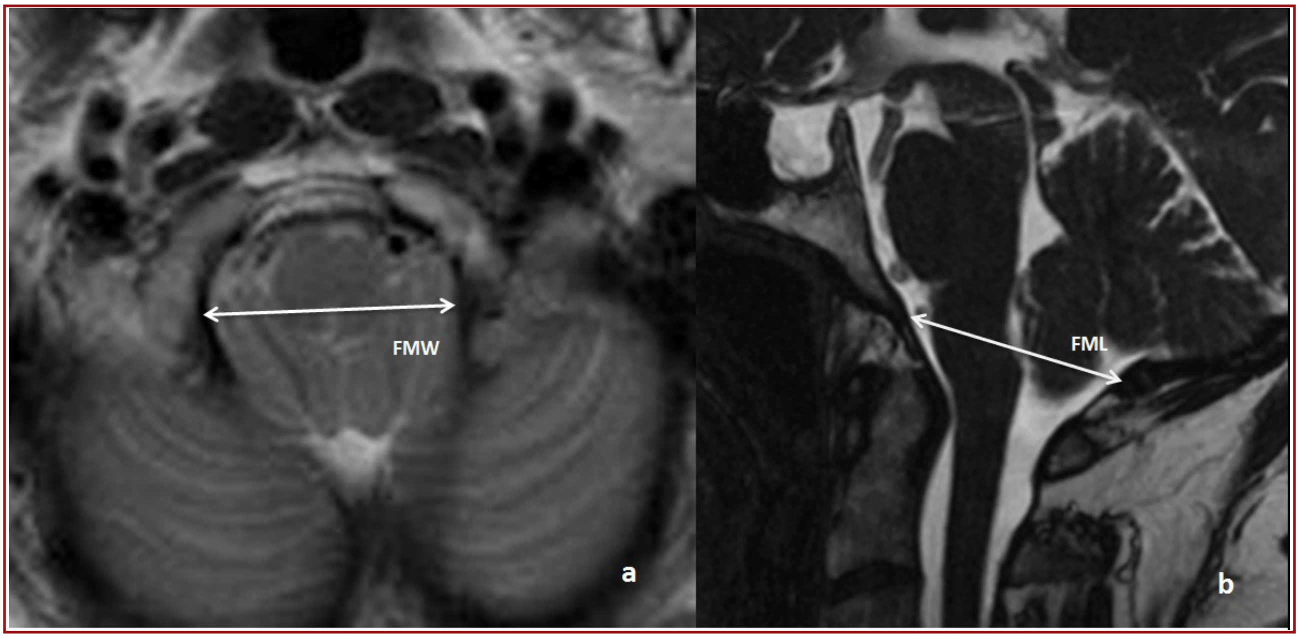

Figure-3. The formula created by Radinsky was used to calculate the Foramen magnum (FM) field.

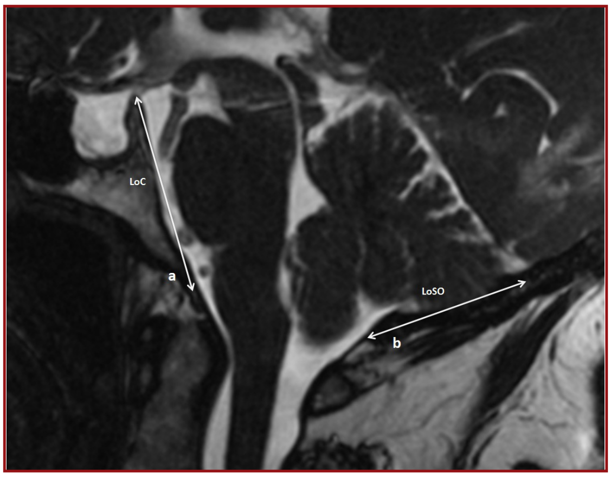

Figure-4. The length of the clivus (LoC), the distance from the top of the dorsum sella to basion; the length of the subocciput (LoSO), the length between the internal occipital protuberance and the opistion

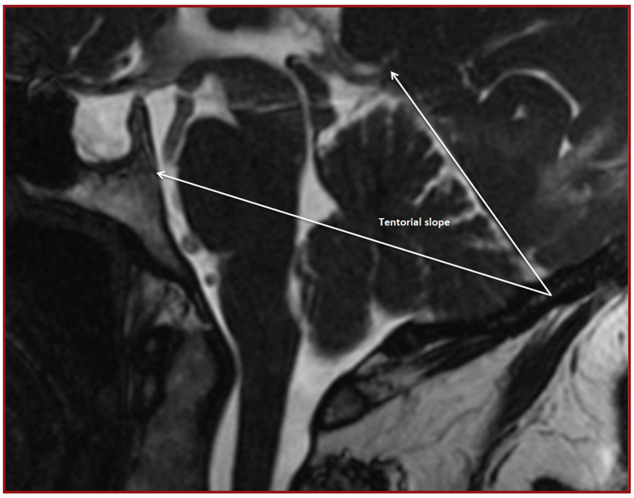

Figure-5. Tentorial slope (TS) was obtained by measuring the angle between the tentorium and the Twining line in the midsagittal cranial MR scan.

Tentorial slope

Tentorial slope (TS) was obtained by measuring the angle between the tentorium and Twining's line on a midsagittal head MR scan (Figure-5).

Clivo-dental angle

Clivo-dental angle (CD angle) is called as Wackenheim clivus base line or cranial-vertebral angle. It is constructed by drawing a line along the clivus and extrapolating inferiorly into upper cervical spinal canal along posterior surface of odontoid bone (Figure-6).

This line should fall tangent to the posterior aspect of the tip of the odontoid process [36].

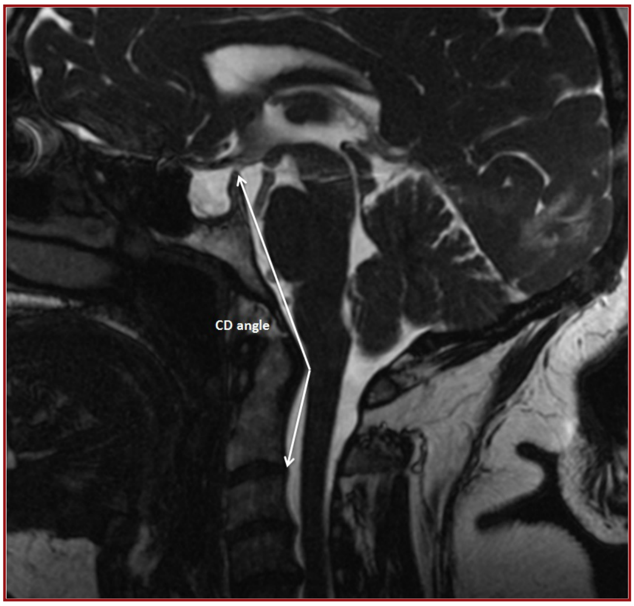

Figure-6. Klivo-Dental Angle (CD Angle) is called Wackenheim Klivus Baseline or Cranial-vertebral Angle. Drawing a line through the clivus and along the posterior surface of the odontoid bone, the upper cervical spinal canal is got at inferiorly. This line should be tangential to the posterior face of the odontoid process tip.

Angle of the clivus and tentorium

The angle between clivus and tentorium (C-T angle) is important for superior compression to structures of posterior fossa. It is constructed by drawing a line along the clivus and another line along the tentorium (Figure-7).

Angle of the clivus and suboccipút

The angle between clivus and sub-occiput (C-SO angle) is like a mouth of cone between the clivus and suboccipút. It is constructed by drawing a line along the clivus and another line along the suboccipút (Figure-8).

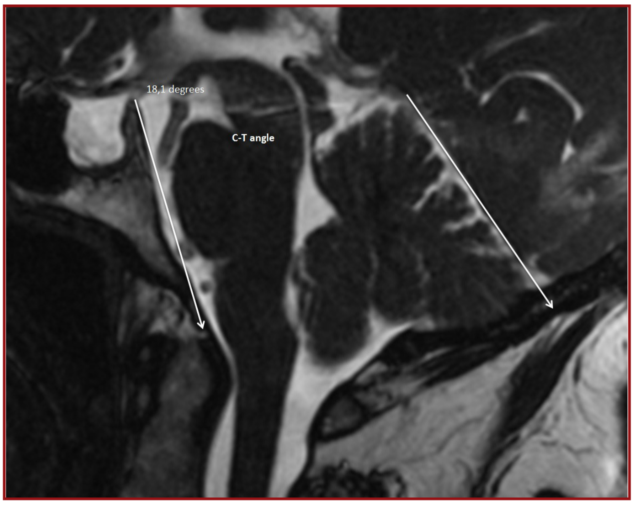

Figure-7. Clivus and Tentorium Angle (C-T angle) is important in the posterior fossa compression superiorly. It was created by drawing a line through clivus and another line through the tentorium.

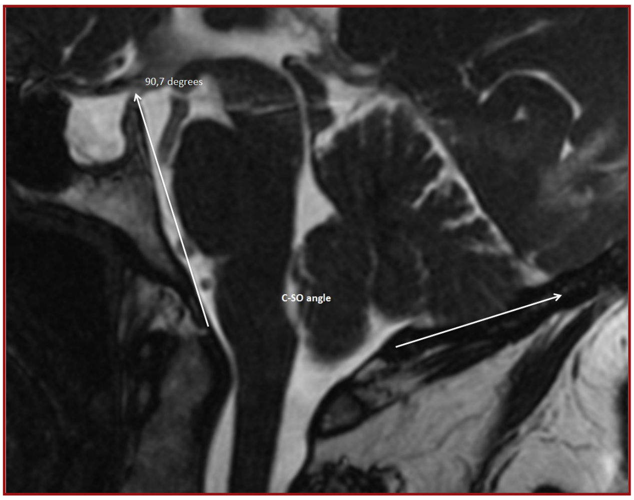

Figure-8. The angle between the clivus and suboxiput (C-SO angle) is similar to the cone mouth. It was created by drawing a line through clivus and another line along the suboccipút

Statistical analysis

The data was collected, tabulated and statistically analyzed. Data was analyzed using IBM SPSS program. Descriptive statistics including range, mean and standard deviation was calculated for each parameter. We assessed test of normality with Kolmogorov- Simirnov test. For normal distribution, we assessed mean differences in dimensions of the posterior fossa, volume of posterior fossa and measurements of the occipital bone for study and control groups using independent-sample Student's t-tests. Significance was indicated by a two-tailed with p value < 0.05, and 95% confidence intervals. For abnormal distribution we used the tests of One way ANOVA, Mann Whitney U and Kruskall Wallis

RESULTS

In our study, 27 cases were evaluated. The cases were 12 patients with CM-1 and 15 ones were control group. The age range of the patient group ranged from 2 to 17 years with a mean age of 12.66 ± 4.41. The age range of the control group ranged from 4 to 17 years, with an average age of 10.40 ± 4.62. Gender distribution in the patient group was 5 male, 7 female; In the control group, sex distribution was 6 males, 9 females. Table 1 summarizes the results between the patient and control group and age and gender in our study (Table-1).

In our study, comparative CD Angle (p: 0,007) and C-SO Angle (p: 0,017) were statistically significant (Table-2).

Table 1. Distribution of age and sex in patient and control groups

		Groups			
		Patient		Control	
		n	Mean±SD	n	Mean±SD
Sex	Male	5		6	
	Female	7		9	
Age			12,66±4,41		10,40±4,62

DISCUSSION

The posterior cranial fossa (PCF) is the most posterior aspect of the skull base and it houses the brainstem, cranial nerves and cerebellum. The basilar, condylar and squamous parts of the occipital bone and the mastoid part of the temporal bone form the floor of the PCF. PCF has a roof formed by the tentorium cerebelli, which is a fold of the dura. The cranial-vertebral (or craniocervical) junction (CVJ) is a collective term that refers to the occiput (posterior skull base}, atlas, axis, and supporting ligaments. PCF has rhomboid shape. The floor of rhomboid is composed of clivus and supraoccipút, the roof is composed of tentorium cerebelli and mesencephalic aperture.

CM is the downward displacement of the caudal part of the cerebellum and/or medulla oblongata into the spinal canal [8]. The pathogenesis of CM is incompletely understood. CM is considered to be a primary neurological disease involving the posterior cranial fossa and the hindbrain [4]. Many investigators have tried to explain the pathogenesis of CM from the standpoint of primary neural anomaly [4,11,21]. However, clinical and experimental studies indicate that the chronic tonsillar herniation observed in CM could result from overcrowding within a primary small and shallow PCF due to an underdeveloped occipital bone [24]. It is produced by a raised intracranial pressure, which has a varied etiology like hydrocephalus, space occupying lesions and a malformed posterior fossa. When the study of Milhorat TH et al was correlated with etiological factors, the following causal mechanisms were suggested: (1) cranial constriction; (2) cranial settling; (3) spinal cord tethering; (4) intracranial hypertension; and (5) intraspinal hypotension [23].

Sgouros S et al. have shown a study of cranial development in healthy children the rate of increase in intracranial volume is not linear throughout the examined period but displays different phases. There is a rapid linear growth during the first 5 years of life. In subsequent years, growth continues but at a much slower rate, with a mild spurt starting at approximately 10 years and lasting for an additional 5 years. This model of intracranial volume growth slightly differs from models based on skull radiographs in both the rate of growth in the first 5 years and the absolute volume values [31].

Table 2. Comparison of volume, length and angle between patient and control groups

	Patient	Control	*p*
	Mean ± SD	**Mean ± SD**	
ICV (cm3)	1137,646±104,646	1104,190±145,972	**0,511**
PFV (cm3)	279,862±38,755	271,028±38,855	**0,562**
FMA (cm2)	9,076±2,369	8,581±1,409	**0,506**
PFV/ICV	0,247±0,039	0,246±0,027	**0,957**
FMA/PFV	0,032±0,006	0,032±0,004	**0,796**
FMA/ICV	0,008±0,002	0,008±0,001	**0,722**
Tentorial slope (TS) (°)	45,683±6,862	48,533±5,767	**0,252**
Clivus length (LoC) (mm)	35,092±5,302	35,327±3,673	**0,893**
Subocciput length (LoSO) (mm)	36,358±4,502	39,020±4,351	**0,132**
Clivus-tentorium Cobb (C-T) angle (°)	14,058±8,122	15,480±7,551	**0,642**
Clivus-subocciput Cobb (C-SO) angle (°)	92,925±6,773	82,793±10,130	**0,007****
Clivodental (C-D) angle (°)	138,408±10,264	147,273±7,670	**0,017***

Independent sample t- test *p < 0,05, ** p < 0,01

There are many studies examining posterior fossa morphology. However, most of them have been done only on CM-1. The study done in the pediatric patient group is very rare.

Studies on CM-1disease; when compared the patient and the control group are examined; In some studies, PFV was found smaller in the patient group [2,3,9,23-24, 32-33,35]. There was no difference between the groups in ISV [3,9,32-33]. In all of the studies, PFV / ISV was found smaller in the patient group [2- 3,9,32-33]. In some studies, LoC [2, 3, 7, 15, 17, 23-25, 32, 37] and LoSO [2-3,6,11,17,23-24,32,37] were found to be smaller in the patient group. To summarize the studies, there is a shrinkage of the posterior fossa, which is led by shortening of the clivus and subocciput dimensions in the patients.

There are no studies showing morphological changes with age because the studies in the literature are made in a certain subgroup of CMs. Sgouros S and colleagues found that the study of intracranial volume in healthy children naturally shows an increase in ISV with age [31].

In children studies, there was no difference between the LoC [6,9,15,25] and LoSO [9,15,25] groups. There was no significant difference between groups in terms of TS [2,17,33] and C-D angle [2]. In further studies, C-D angle was narrower in the patient group [15,37]. Again, the TS in CM-1 patients in a study conducted in Turkey were found to be larger [17]. Furtado SV and colleagues have shown that the FM field is compatible with a specific ISV in pediatric CM-1 patients and that both the ISV and FM area are not significantly different from the normal pediatric population [10]. Furtado and colleagues did not detect any differences in morphological value with respect to the mean age in pediatric and adult CM-1 cases [9]. Trigylidas and colleagues compared 0 to 9 years of age and 10 to 18 years of age in their study of children under 18 years of age. In the patient group, PFV / ISV ratio with age showed a significant change in the 0-9 age range, whereas in the 10-18 age group, there was no difference with the control group [33]. To summarize the studies performed in pediatric CM cases, there is no difference in volume, length and angle measurements between healthy individuals.

There was no difference in volume studies in our study. Only C-D Angle is found to be narrower in CM patients. This means increased pressure in the anteroinferior of the brain stem. Because of the steep transition between the cranium and the spinal canal, brain stem findings can be seen in patients. In addition, we found a larger angle between clivus and subocciput in patients. İn the sagittal examination with posterior fossa, clivus and subocciput; the tentorium and the tentorial diaphragm form the rhomboid superiorly. Our findings show that the base of this rhomboid region is larger in patients; suggesting that the structures in the posterior fossa have a larger base.

CONCLUSION

This study showed no difference between intracranial volumes and cranial-vertebral junction length measurements in Pediatric Type 1 Chiari Malformation according to the healthy control group. Again, there is a steep transition between the clivus and the dens as in adult CM patients in the cranial-vertebral compartment, which leads to brain stem findings. It should also be noted that the cerebellum is on a wider surface in CM patients.

Conflict of Interest

There is no conflict of interest.

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