MORPHOMETRIC ANALYSIS OF POSTERIOR CRANIAL FOSSA IN HEALTHY ADULT POPULATION AND ADULTS WITH CHIARI 1 MALFORMATION

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CERTIFICATE

This is to certify that this dissertation entitled **"MORPHOMETRIC ANALYSIS OF POSTERIOR CRANIAL FOSSA IN HEALTHY ADULT POPULATION AND ADULTS WITH CHIARI 1 MALFORMATION"** submitted by **Dr. B. Rohit** appearing for **M.Ch.** Degree examination in **August 2010** is a bonafide record of work done by him under my direct guidance and supervision in partial fulfillment of regulations of the Tamil Nadu Dr. M.G.R. Medical University, Chennai. I forward this to the Tamil Nadu Dr.M.G.R. Medical University, Chennai, Tamil Nadu, India.

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DECLARATION

I **Dr. B. Rohit,** do solemnly affirm that this dissertation titled **"MORPHOMETRIC ANALYSIS OF POSTERIOR CRANIAL FOSSA IN HEALTHY ADULT POPULATION AND ADULTS WITH CHIARI 1 MALFORMATION"** is done by me at Institute of Neurology, Madras Medical College & Govt. General Hospital, Chennai, during 2008-2010 under the guidance and supervision of **Prof. V. Sundar M.Ch. Professor of Neurosurgery, Institute of Neurology.**

The dissertation is submitted to The Tamilnadu Dr. M.G.R. Medical University towards the partial fulfillment of requirements for the award of **M.Ch, Degree in Neurosurgery.**

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CONTENTS

INTRODUCTION

Chiari malformation is a hindbrain malformation characterized by a downward herniation of the caudal part of the cerebellum and/or medulla oblongata into the spinal canal. More cases of Chiari 1 malformation(CM-I) are being diagnosed recently with the advent of magnetic resonance imaging(MRI) of brain. Syringomyelia is a common finding in CM-I.

Chiari malformation is considered to be a primary neurological disease involving the posterior cranial fossa(PCF) and the hindbrain[5].The exact pathogenesis of CM-I is not clear. Many investigators have tried to explain the pathogenesis of Chiari malformation from the standpoint of primary neural anomaly [5,18,29].

Experimental models[26,27,34] supported by morphometric studies[4,31,32,39,40,42] have shown that the chronic tonsillar herniation(CTH) occurring in classic CM-I mainly results from overcrowding of a normally developing hindbrain within a congenitally small and shallow posterior cranial fossa(PCF) due to occipital bone underdevelopment.

Recent morphometric studies focussing on the bony part of the posterior cranial fossa in adult patients with Chiari malformation have lent support to this hypothesis[39,42,45] .

There has been no morphometric study of the PCF from the Indian adult population.This study is an attempt to analyse the morphology and dimensions of the bony and neural structures of the PCF in the Indian adult population and to study the correlation between the analysed parameters and tonsillar herniation.

AIM

1. To estimate the volume and bony dimensions of the posterior cranial fossa in healthy adult population and adult patients with Chiari 1 malformation.

2. To study the morphology of the neural structures(brainstem and cerebellum) within the posterior cranial fossa in healthy adult population and adult patients with Chiari 1 malformation .

REVIEW OF LITERATURE

The literature on Chiari malformation is reviewed under the following aspects

1. CLASSIFICATION OF HIND BRAIN MALFORMATIONS
2. TYPES OF CHIARI MALFORMATION
3. CHIARI 1 MALFORMATION
    - FEATURES
    - SYMPTOMS AND SIGNS
    - IMAGING
    - MANAGEMENT
4. STUDIES ON MORPHOLOGY OF POSTERIOR CRANIAL FOSSA IN NORMAL POPULATION AND CHIARI I MALFORMATION.

Hindbrain malformations can be classified as given in the table below.

CLASSIFICATION SCHEME FOR MALFORMATIONS OF HINDBRAIN DEVELOPMENT[30]

Malformations of both midbrain and hindbrain	Malformations affecting predominantly the cerebellum and derivatives	Malformations affecting predominantly the lower hindbrain	Posterior fossa abnormalities	Malformations associated with prenatal onset degeneration
Brainstem-cerebellar hypoplasia-dysplasia Chiari II malformations Cobblestone LIS with mid-hindbrain malformation Molar tooth sign associated malformations Joubert syndrome JSRD, including Senior–Loken and COACH Rhombencephalosynapsis	Focal cerebellar hypoplasia (focal or hemispheric) Paleocerebellar hypoplasia (vermis predominantly affected, brainstem often mildly hypoplastic) Dandy–Walker malformation Cerebellar vermis hypoplasia, isolated CVH with periventricular nodular heterotopia CVH with cortical malformations (LIS, PMG) Neocerebellar hypoplasia (hemispheres and vermis affected, predominantly granule cell hypoplasia)	Chiari I malformations Cranial nerve and nuclear aplasias Moebius syndrome Duane retraction syndrome	Abnormal fluid collections Arachnoid cyst Blake's pouch cyst Mega-cisterna magna Abnormal bone and brain structure	Ponto-cerebellar hypoplasia (PCH) PCH type 1, PCH type 2, PCH type 3 Congenital disorders of glycosylation (CDG)

[CDG - congenital disorders of glycosylation; COACH - Cerebellar vermis hypoplasia, Oligophrenia, Ataxia, Coloboma, and Hepatic fibrosis;CVH - cerebellar vermis hypoplasia; JSRD - Joubert syndrome and related disorders; LIS - lissencephaly; PMG - polymicrogyria]

Between 1891-1896,Hans Chiari an Austrian pathologist described a series of anomalies of the caudal cerebellum and brainstem on the basis of autopsy observations and published his paper *Ueber Veränderungen des Kleinhirns infolge von Hydrocephalie des Grosshirns* (Concerning alterations in the cerebellum resulting from cerebral hydrocephalus), in1896 on the Chiari malformations.

In 1894, Julius Arnold a German pathologist also described this disorder,in a myelodysplastic patient and published his paper titled *Myelocyste, Transposition von Gewebskeimen und Sympodie***.**

In 1907, two of Arnold's students Schwalbe and Gredig**,** coined the eponym "Arnold-Chiari malformation" to Chiari Type II malformation in honor of both men.

CHIARI MALFORMATION - TYPES

I	:	Caudal displacement of cerbellar tonsil > 5 mm below the plane of foramen magnum
II	:	Caudal displacement of cerebellar vermis, fourth ventricle, lower brainstem below the plane of foramen magnum , commonly associated with myelodysplasia.
III	:	Caudal displacement of the cerebellum and brainstem into a high cervical meningocele
IV	:	Cerebellar hypoplasia
0	:	Cerebrospinal fluid (CSF) equilibrium changes at craniocervical junction.

Syringohydromyelia present

Absent hindbrain herniation

1 ½ : Tonsillar herniation

Elongated brain stem and fourth ventricle

No intracranial abnormalities

Various theories were proposed to explain the hindbrain findings of Chiari malformations.

- Gardner advocated the hydrodynamic theory and believed that hydrocephalus and hydromyelia were normal physiologic events in early embryologic development.The hydrodynamic theory, however, did not explain the small size of the posterior fossa, the upward herniation of the posterior fossa contents, the slit-like fourth ventricle and the multiple supratentorial anomalies.

- The traction theory was proposed by Penfield, Cobum and Luchenstein, who suggested that a tethered spinal cord near the myelomeningocele may pull the cerebellum and medulla into the cervical canal. In the Chiari II malformation, however, the spinal cord is not always tethered. Traction from the caudal end of the cord is rapidly dissipated within 4 segments. The theory did not explain the medullary kink.

- Cleland proposed the developmental arrest theory with primary dysgenesis of the brainstem as the main cause of the malformation.

- Daniel, Stritch, and Peach thought that failure of development of the pontine flexure due to a developmental arrest causes both upward and downward herniation of the elongated brainstem. This theory did not explain the associated cerebral malformations.

CHIARI 1 MALFORMATION

Chiari I malformation (CM-I) is characterized by:

- Herniation of the cerebellar tonsils more than 5 mm through the foramen magnum into the cervical spinal canal.
- The cerebellar tonsils often are elongated.
- Mild caudal displacement and flattening or kinking of the medulla may be present.

The vermis cerebelli and the fourth ventricle are normal or only minimally deformed.

CM-I may have a genetic basis, as suggested by:

- Its association with known genetic disorders such as achondroplasia, Hadju-Cheney syndrome, and Klippel-Feil syndrome.
- Reports of familial aggregation and concordance among monozygotic twins and triplets.
- Findings from pedigree analyses in some families suggest an autosomal dominant inheritance pattern with reduced penetrance or an autosomal recessive inheritance pattern.

Acquired CM-I is reported to develop after lumboperitoneal or ventriculoperitoneal shunt placement.

Clinical Features

- Suboccipital headache [23].

- Ocular symptoms, including retro-orbital pain, visual disturbances, photophobia, and diplopia[25,28,41].

- Otoneurologic symptoms, including dizziness, vertigo, hearing disturbances, oscillopsia, nystagmus[6,19,20,22].

- Hindbrain compression symptoms, including weakness, paresthesia, ataxia, cranial nerve palsies, dysphagia, dysphasia, palpitations, syncope, apnea, and sudden death[1,2,3,9,12].

- Syringomyelia symptoms, including central cord syndrome, impaired sensation, impaired motor control, gait disturbance, torticollis, bowel and/or bladder symptoms [12,17,31,32].

- Patients who have CM-I and a syrinx almost always present with symptoms referable to the syrinx.

- If the syrinx extends into the medulla, bulbar symptoms predominate.

- In pediatric patients, the most common symptoms are headache, neck pain, and ataxia.

- The natural history of CM-I is not understood clearly. Many patients are asymptomatic and probably remain so all of their lives.

Imaging:

Radiograph

Osseous anomalies – seen in 25-50% of pts with CM-I:

Basilar invagination (25-50%)

- Atlantooccipital assimilation (1-5%)
- Klippel-Feil syndrome (5-10%)
- Incomplete ossification of C1 ring (5%)
- Proatlantal remnant spina bifida at the C1 level
- Retroflexed odontoid process (26%)
- Scoliosis (42%)
- Kyphosis
- Increased cervical lordosis
- Cervical ribs
- Fused thoracic ribs

Computed Tomography(CT) Scan

- Obliterated cisterna magna
- Hydrocephalus
- Flattened spinal cord
- Tonsillar ectopia

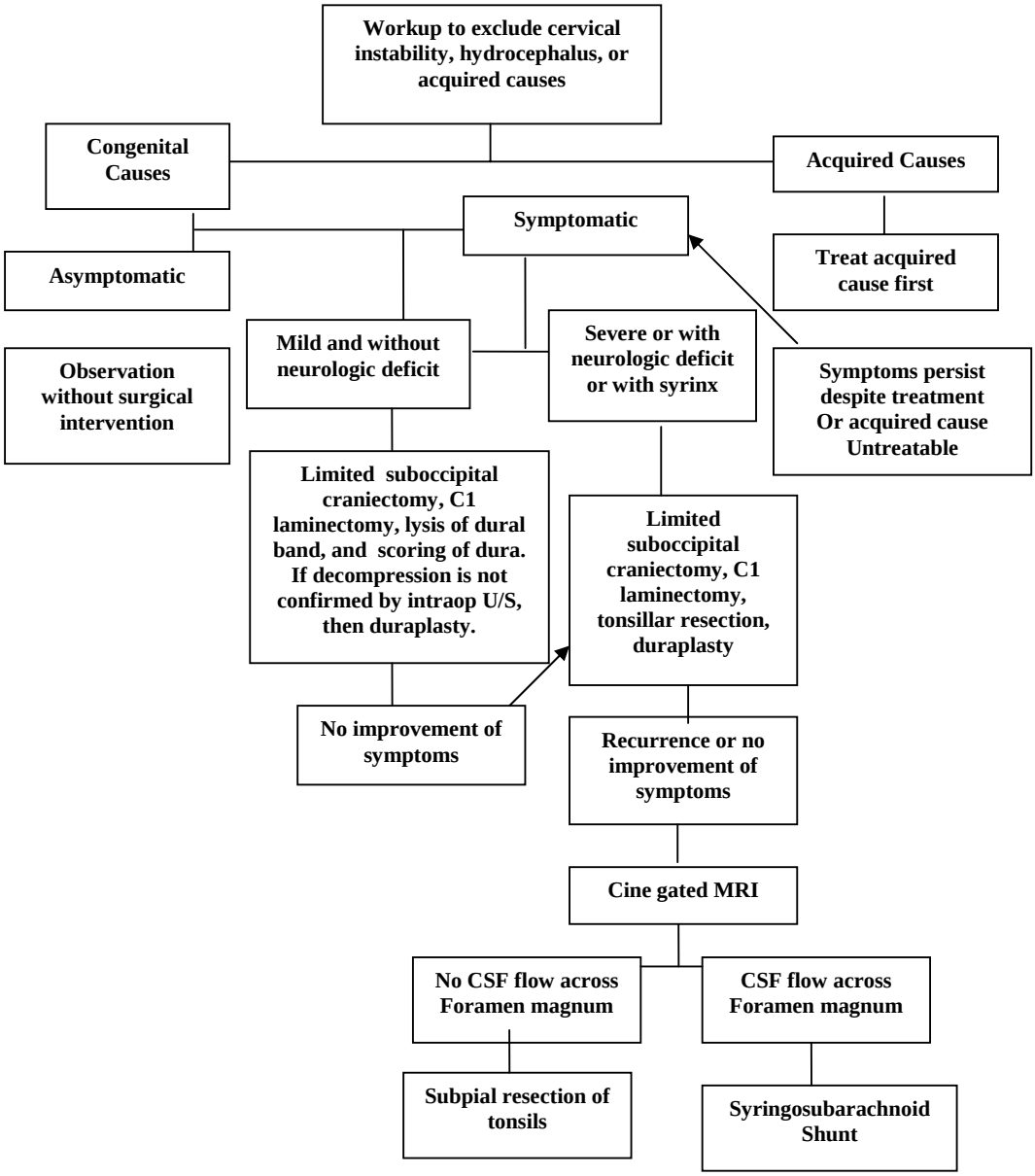
MRI Scan

- Displacement of cerebellar tonsils below the level of the foramen magnum
- Pointed tonsils
- Elongation of the fourth ventricle
- Hindbrain abnormalities
    - Obstructive hydrocephalus
    - Associated abnormalities such as syringomyelia.

The spinal cord cavity fills from the spinal subarachnoid space as a consequence of CSF waves produced by the piston-like effect of impacted cerebellar tonsils driving CSF through the extracellular spaces of the spinal cord into the central canal.

MANAGEMENT

The management protocol for patients with CM-I is depicted in the flowchart below



STUDIES ON MORPHOMETRIC ANALYSIS OF PCF

The posterior fossa is defined as the osseous anatomical area with a floor formed by the occipital bone (basiocciput portion of the clivus and

supraocciput portion of the occipital bone up to the insertion of the tentorium cerebelli, which forms the superior boundary of this fossa) and the basisphenoid. The petrous ridges of the temporal bones form the anterolateral border of this cavity anteriorly to their connection (posterior petroclinoid ligament) to the posterior clinoids.

Tubbs et al (2010)[44] have performed an anatomical study of the posterior cranial fossa.To elucidate the anatomic variations in the morphology of the foramen magnum the authors conducted a morphometric study using dry skulls . Digital images were obtained of the foramen magnum from an inferior view. These images were studied using a computer-assisted image analysis system and an image processor was used to calculate pixel differences between 2 selected points, which allowed accurate translation of pixel differences into metric measurements. The mean surface area of the foramen magnum was 558 mm^2, the mean anteroposterior diameter was 3.1 cm, and the mean horizontal diameter was 2.7 cm. For comparison, surface areas were classified into 3 types based on size. Type I foramina were identified in 20.8% of the dry skulls (15 skulls) and exhibited a surface area of less than 500 mm^2. Type II (66.6%, 48 skulls) was applied to foramina of an intermediate size with surface areas ranging between 500 to 600 mm^2. Type III (12.5%, 9 skulls) was applied to large foramina with surface areas of more than 600 mm^2.

There have been various studies to elucidate the pathogenesis of Chiari 1 malformation

Two neuropathologists, Uros Roessmann and Reinhart Friede (1976)[16] studied all cases of CTH that had been published earlier and postulated that all cases of herniated cerebellar tonsils were related to distortion of the brainstem and cerebellum at the foramen magnum from multiple causes:brain distortion from above, as in the case of multiple sutural closure in syndromic craniosynostosis; suctioning of the brain from below, as in response to lumboperitoneal shunting or direct distortion by bony abnormalities of the craniovertebral junction, as in basilar invagination.

Marin Padilla – Marin Padilla et al(1981,1991) [26,27] postulated that CM-I is a disorder of paraxial mesoderm of parachordal plate that leads to occipital bone underdevelopment and a small posterior cranial fossa which is overoccupied and then over filled during post natal growth spurt of cerebellum, which is found to grow in the available space of upper cervical canal.Caudal displacement of brainstem can enlarge the foramen magnum and simultaneously cause upward shift of the cerebellar tentorium. Symptomatic CM-I is a dynamic disease and the number of patients increases with age. Tardive closure of spheno occipital suture growing zone occurs not only in embryological period but also in infancy.Because of its early development , basiocciput hypoplasia could affect the subsequent development of every cranial bone of cephalic region. Posteriorly it articulates with exoccpital bones and could determine the location and orientation of PCF as well as the position of temporal bones laterally , orofacial and pharyngo laryngeal cavities

anteriorly. CM-I is associated with a subtype of orofacial skeletal malformation adapting short and lordotic axial basicranium.

Vega et al(1990)[45] determined the frequency of anomalies of the basichondrocranium in a series of 42 patients with Chiari type I malformation compared with a control group of 46 subjects. Sixteen patients had syringomyelia. Linear, angular and posterior fossa surface area measurements were taken on conventional lateral skull x-rays. Posterior fossa volume was estimated by CT scanning. In patients there was shortening of clivus length, twining-opisthion distance and chamberlain's line. The size of the posterior fossa was smaller in patients than in controls. The most discriminative variables were posterior fossa area and clivus length which allowed accurate identification of 76% of patients as belonging to the patient group and 79% of controls as belonging to the control group. The authors suggested that under-development of the basichondrocranium with a small size of the posterior fossa is an outstanding feature in adult Chiari type I malformation and hypothesised that tonsillar ectopia is secondary to the disproportion between the posterior fossa and the cerebellum, which is forced to grow into the cervical spinal canal.

Stovner et al(1993) [42] measured skull dimensions in 33 adult patients with MRI-verified Chiari I malformations and in 40 controls. The posterior cranial fossa was significantly smaller and shallower in patients than in controls.

To investigate overcrowding in the posterior cranial fossa as the pathogenesis of adult-type Chiari malformation, Nishikawa et al(1997) [33] studied the morphology of the brainstem and cerebellum within the posterior cranial fossa (neural structures consisting of the midbrain, pons, cerebellum, and medulla oblongata) as well as the base of the skull. Thirty patients with Chiari malformation and 50 normal control subjects were prospectively studied using neuroimaging. To estimate overcrowding, the authors used a "volume ratio" in which volume of the posterior fossa brain (consisting of the midbrain, pons, cerebellum, and medulla oblongata within the posterior cranial fossa) was placed in a ratio with the volume of the posterior fossa cranium encircled by bony and tentorial structures. Compared to the control group, in the chiari group there was a significantly larger volume ratio, the two occipital enchondral parts (the exocciput and supraocciput) were significantly smaller and the tentorium was pronouncedly steeper. There was no significant difference in the posterior fossa brain volume or in the axial lengths of the hindbrain (the brainstem and cerebellum). In six patients with basilar invagination the medulla oblongata was herniated, all three occipital enchondral parts (the basiocciput, exocciput, and supraocciput) were significantly smaller than in the control group and the volume ratio was significantly larger than that in the Chiari group without basilar invagination. They suggested that in adult-type Chiari malformation an underdeveloped occipital bone, possibly due to underdevelopment of the occipital somite originating from the paraxial mesoderm, induces overcrowding in the posterior

cranial fossa, which contains the normally developed hindbrain. Basilar invagination is associated with a more severe downward herniation of the hindbrain due to the more severely underdeveloped occipital enchondrium, which further exacerbates overcrowding of the posterior cranial fossa.

The term "classic CM-I," was introduced by Milhorat(1999)[31] to describe cases of CM-I related to an isolated constriction of the PCF. This term depicts a condition in which there is no other inductive cause of tonsillar descent such as multiple faciocraniosynostosis, hydrocephalus, lumboperitoneal shunting or intracranial hypertension. Milhorat analysed the posterior cranial fossa volume using cavalieri method in patients with CM-I and age specific controls and found out that posterior cranial fossa volume is decreased in CM-I but not in control group.

Aydin_ et al(2005) [4] examined a retrospective cohort of 60 adult patients with CM-I and multiple measurements were made on magnetic resonance imaging and the results compared to 30 healthy adult control subjects. All measurements except mean anteroposterior diameter of the foramen magnum were reduced in patients compared to controls. An increase in the anteroposterior mid-sagittal distance of the foramen magnum in patients reached statistically significant difference compared to controls. This study suggested that the bony components of the PCF are not developed fully, supporting the current concept that CM-I is a disorder of the para-axial mesoderm.

In order to establish smaller geometry of PCF as the cause of hindbrain herniation in a family,Tubbs et al(2008)[37] performed volumetric analysis in a family found to have Chiari 1 malformation documented in 4 generations. Members from the family found to have a CM-I by imaging underwent volumetric analysis of their posterior cranial fossa using the Cavalieri method. No member of the family found to have CM-I on preoperative imaging had a posterior fossa that was significantly smaller than that of age-matched controls.They postulated that not all patients with a CM-I will have a reduced posterior cranial fossa volume and although the mechanism for the development of hindbrain herniation in this cohort is unknown, this manifestation can be seen in multiple generations of a familial aggregation with normal posterior fossa capacity.

Noudel et al(2009) [38] assessed the relationship between the descent of the cerebellar tonsils and dimensions of the basiocciput, specifically the length of the clivus. They studied 17 patients who had symptoms and signs related to the descent of cerebellar tonsils more than 5 mm below the foramen magnum. The patients were all symptomatic and all had a herniation of at least 5 mm.They proposed that a cranioencephalic disproportion between the normal cerebellum and a small PCF, even when restricted to the basioccipital portion, is a sufficient condition to explain the development of a symptomatic CM-I. The authors hypothesized that the predominance of basioccipital hypoplasia could proceed either from an early paraxial mesodermal insufficiency due to a

failure of notochordal induction in utero or from a premature stenosis of the sphenooccipital synchondrosis occurring later in infancy. This theory explains that congenital or very early symptomatic CM-I is probably related to a true abnormal formation of the occipital bone precursor during the intrauterine life and later CM-Is is caused by a sphenooccipital synchondrosial growth anomaly. The authors proposed that basioccipital measurement could help in establishing the diagnosis of classical CM-I. The demonstration of basioccipital hypoplasia could be a means of differentiating true cases of CM-I from incidentally diagnosed cases. Cases of CM-I related to basioccipital hypoplasia may represent a specific subtype in which orofacial skeletal malformations resulting from the adaptation of the facial skeleton to a primarily short and lordotic axial basicranium are more frequently observed.

Hegde et al(2010) [43] studied foramen magnum dimensions and intracranial volume in Chiari I malformations in Indian children.The area of the foramen magnum was obtained independently using computer imaging software and a regression formula. The result of 21 pediatric patients was compared with a matched control group. Though the volume was significantly reduced in the Chiari group, there was no statistical difference in the area and linear dimensions of the foramen magnum in the study and control groups. The authors concluded that irregular shape of the foramen magnum is accentuated by developmental bony and soft tissue anomalies at the cranio-vertebral junction in Chiari malformation.

MATERIALS AND METHODS

In this study an analysis of the morphometry of posterior cranial fossa in 30 patients with Chiari I malformation who were symptomatic and 44 healthy controls was done.

All patients who were referred to the Institute of Neurology between August 2007 and March 2010 were enrolled in the study. The control group included 44 patients who underwent brain MR imaging for headaches or migraine in the same period and whose MRI was normal.Control group included patients aged more than 16 years and were enrolled from the outpatient section and the radiology department.

Inclusion criteria (CM-I)

- Age > 16 Years
- Tonsillar descent of ≥ 5mm on midsagittal MRI

Exclusion criteria

- Patients with hydrocephalus (operated), who have undergone thecoperitoneal stunt, basilar impression, craniosynostosis.

METHODS

This study compared the parameters between 30 adult patients with CM-I and 44 healthy subjects. The patient group consisted of 11 women and 19 men, with a mean age of 30 years. The control group consisted of 33 women and 11 men, with a mean age of 36 years.

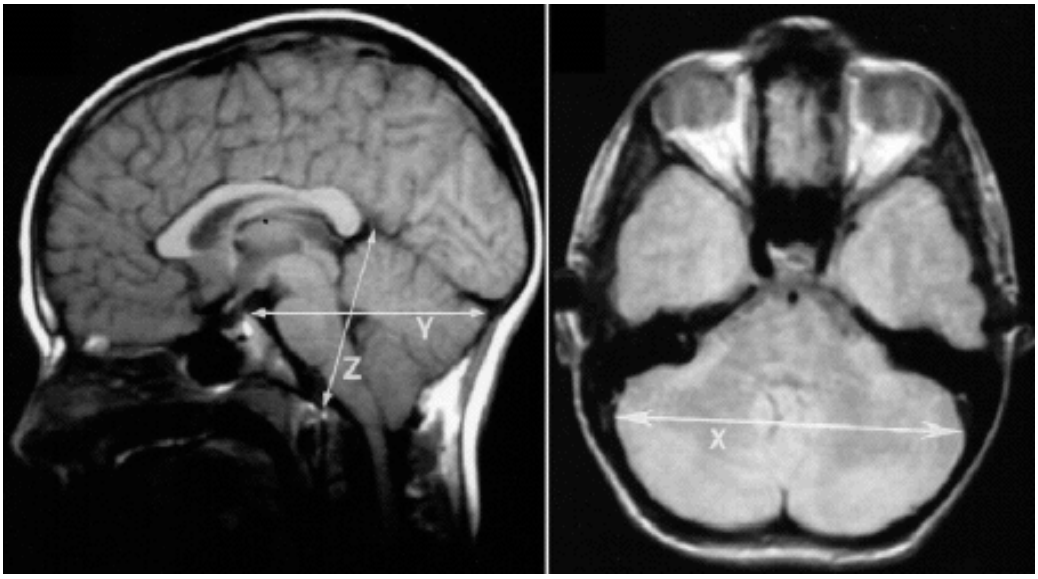
Twelve patients had CM-I only (40%), whereas eighteen patients had CM-I with syringomyelia (60%). Eleven patients presented with a history of paroxysmal occipital headaches,11 patients with syringomyelia had radicular pains ,wasting and thermoalgesic dissociation, 4 patients displayed bipyramidal signs and 4 patients had cerebellar symptoms and signs.

Using midsagittal MR imaging, the following linear measurements of the PCF were made

x - maximum width of bony posterior cranial fossa

y - distance from posterior clinoid process to torcula heterophili

z - Height of Posterior Cranial fossa - from basion to peak of tentorium cerebelli



The volume of the posterior cranial fossa was calculated using the formula,

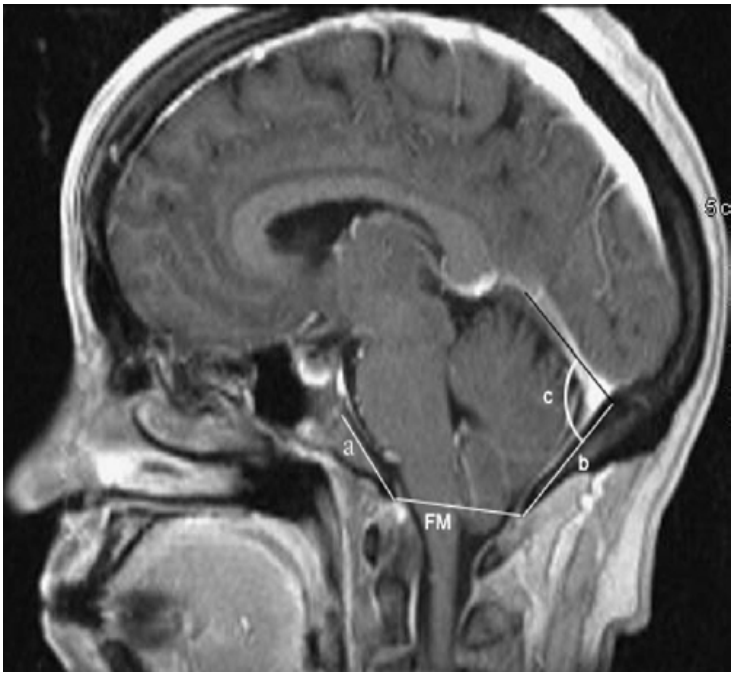
PCF Volume - 4/3 π x **(x/2** x **y/2** x **z/2)**

The volume of PCF in the controls was calculated in a similar fashion.

The length of the basiocciput (**a**) was measured from the basioccipital synchondrosis to the basion.The basioccipital synchondrosis was clearly visualized in both the groups.

The length of supraocciput (**b**) was measured from the internal occipital protuberance to opisthion.

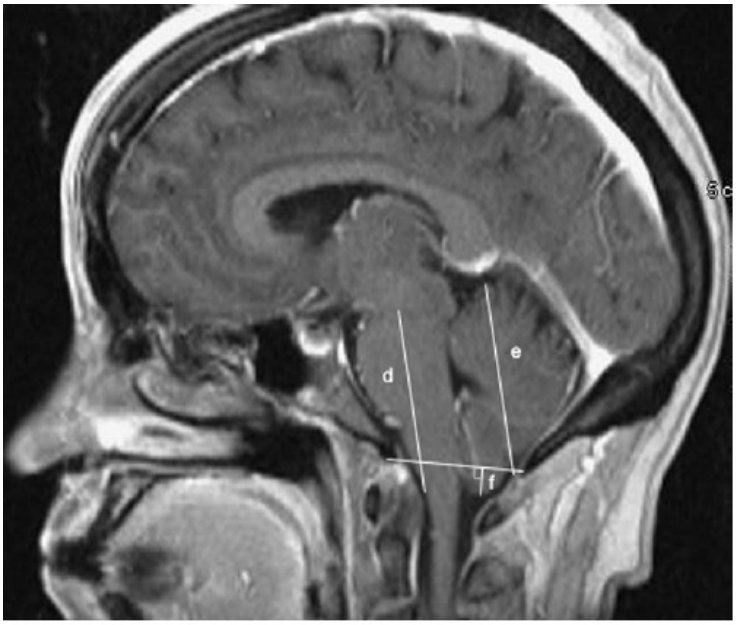
The tentorial angle (**c**) was the angle subtended by a line, connecting the internal occipital protuberance to opisthion, with the tentorium.



The brainstem length (**d**) was calculated from midbrain-pons junction to cervicomedullary junction.

The cerebellar length (**e**) was the length of the line connecting the most rostral and caudal points of the cerebellar hemisphere along a straight line drawn caudally and parallel to the bottom of the fourth ventricle.

The length of tonsillar descent (**f**)was measured by a perpendicular line from the McRae's line to the tip of the tonsil.



Compression of the CSF cisterns posterior and lateral to the cerebellum was identified by the absence of the hyperintense signal at the lowest point of the cerebellar hemisphere on sagittal T2-weighted MR images.

The accuracy of the formula was ascertained using 10 dry skulls, the posterior cranial fossa volume of which was calculated by filling it with sand. A CT was obtained of the dry skull and the above said linear measurements made and the volume calculated using the given formula. There was no statistically significant difference in the volume calculated by either means.

Statistical analysis was performed with the aid of SPSS software for Windows (version 11, SPSS, Inc). The mean differences in the linear measurements and volume of the PCF for controls and Chiari group was measured using independent-sample student's *t* tests. Significance was indicated by a two-tailed P value of less than 0.05.

RESULTS

The details of the morphometric evaluation are given in the Appendix 2.

The mean values of the eight parameters measured for each group are shown in Table 1.

Table 1

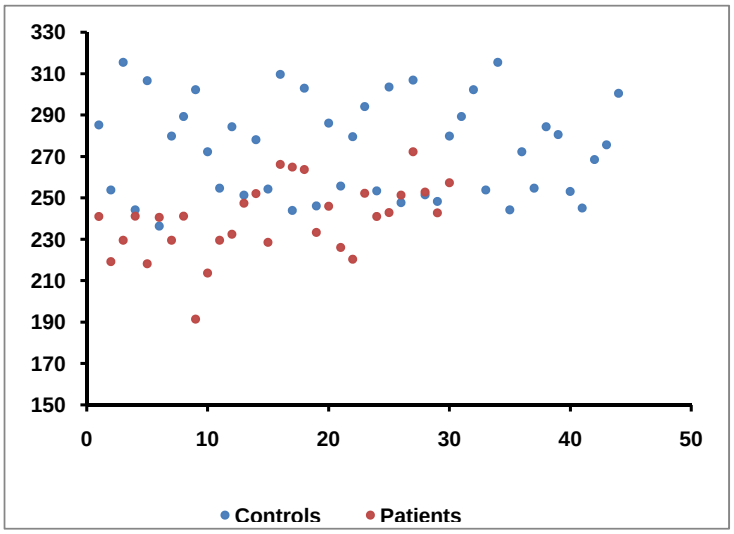
	Group Statistics				
	GROUP	**N**	**Mean**	**Std. Deviation**	**Std. Error Mean**
VOLUME	Control	44	274.03	23.44	3.53
	Chiari	30	239.65	17.61	3.22
BASIOCCIPUT	Control	44	2.73	0.40	0.06
	Chiari	30	2.69	0.31	0.06
FORAMEN MAGNUM	Control	44	3.41	0.33	0.05
	Chiari	30	3.45	0.27	0.05
SUPRAOCCIPUT	Control	44	4.08	0.12	0.02
	Chiari	30	4.09	0.13	0.02
TENTORIAL ANGLE	Control	44	84.32	0.85	0.13
	Chiari	30	85.11	1.11	0.20
BRAINSTEM LENGTH	Control	44	4.87	0.16	0.02
	Chiari	30	4.93	0.15	0.03
CEREBELLAR LENGTH	Control	44	4.94	0.14	0.02
	Chiari	30	4.91	0.13	0.02
HEIGHT	Control	44	7.13	0.33	0.05
	Chiari	30	6.58	0.30	0.05

Independent Samples Test								
		Levene's Test for Equality of Variances		t-test for Equality of Means				
							95% Confidence Interval of the Difference	
		F	Sig.	T	Df	Sig. (2-tailed)	Lower	Upper
Volume	Equal variances assumed	6.853	0.011	6.822	72.000	0.000	24.333	44.426
	Equal variances not assumed			7.196	71.257	0.000	24.854	43.905
Basiocciput	Equal variances assumed	2.429	0.124	0.554	72.000	0.581	-0.125	0.221
	Equal variances not assumed			0.579	70.320	0.564	-0.118	0.214
Foramen magnum	Equal variances assumed	0.050	0.823	-0.506	72.000	0.615	-0.181	0.108
	Equal variances not assumed			-0.526	69.746	0.600	-0.176	0.102
Supraocciput	Equal variances assumed	0.591	0.445	-0.304	72.000	0.762	-0.066	0.049
	Equal variances not assumed			-0.299	58.607	0.766	-0.068	0.050
Tentorial angle	Equal variances assumed	4.057	0.048	-3.441	72.000	0.001	-1.238	-0.330
	Equal variances not assumed			-3.269	51.058	0.002	-1.265	-0.303
Brainstem length	Equal variances assumed	0.035	0.853	-1.736	72.000	0.087	-0.134	0.009
	Equal variances not assumed			-1.758	65.173	0.083	-0.133	0.008
Cerebellar length	Equal variances assumed	0.324	0.571	1.011	72.000	0.315	-0.031	0.095
	Equal variances not assumed			1.030	66.270	0.307	-0.030	0.093
Height	Equal variances assumed	0.804	0.373	7.290	72.000	0.000	0.399	0.699
	Equal variances not assumed			7.416	65.976	0.000	0.401	0.697

Table 2 summarizes statistical results of the parameters evaluated in this study

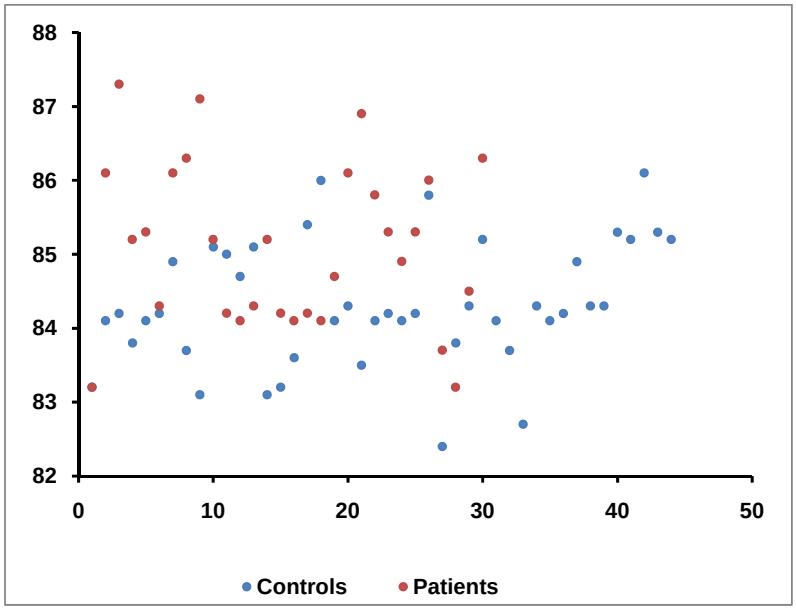
THE SCATTER CHARTS FOR THE STATISTICALLY SIGINIFICANT PARAMETERS ARE DEPICTED BELOW

Scatter chart – VOLUME – Controls and Chiari Group



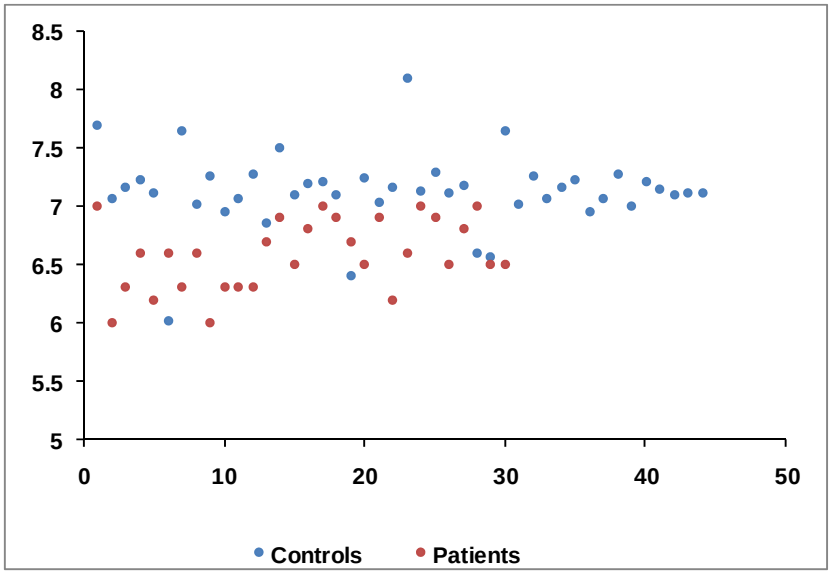
In the 30 patients in whom these data were measured, the mean volume was 239 ±17 cc whereas in the 44 control participants the average volume was 274 ± 23 cc.

Scatter chart – HEIGHT – Controls and Chiari Group



There was no significant difference in the width of the posterior cranial fossa or the posterior clinoid - torcular herophili distance; However, the height of the PCF(basion-peak of the tentorium cerebella) was significantly shorter in those with a Chiari malformation (6.58 ± 0.3 cm) as compared with controls (7.13 ± 0.33 cm; $p < 0.001$).

Scatter chart – TENTORIAL ANGLE – Controls and Chiari Group



Compared to the control group, there was a significant difference (p - 0.001) in the mean angle of the cerebellar tentorium against Twining's line, which was 85.11^0 in the Chiari group and 84.32^0 in the control group. This indicated that the cerebellar tentorium was significantly steeper in the Chiari group than in the control group.

Although the mean length of the Basiocciput in the Chiari group was shorter than that in the control group (2.69 cm in the Chiari group and 2.73 cm in the control group), it was not statistically significant ($p > 0.05$).

No significant difference ($p > 0.05$)was found in the length of the supraocciput (between the internal occipital protuberance and the opisthion), which measured 40.9 mm(mean) in the Chiari group and 40.8 mm(mean) in the control group.

The volume of the posterior cranial fossa was significantly smaller in patients with a Chiari malformation as compared with normal controls ($p < 0.001$).

Comparing the Chiari group to the control group, no significant difference was found in the length of the brainstem from the midbrain-pons junction to the cervicomedullary junction (mean - 4.93 cm in the Chiari group and 4.87 cm in the control group) or in the long axial length of the cerebellar hemisphere (mean - 4.91 cm in the Chiari group and 4.94 cm in the control group).

Overall, compression of the retrocerebellar CSF spaces of the cisterna magna was seen in 100% of our patients.

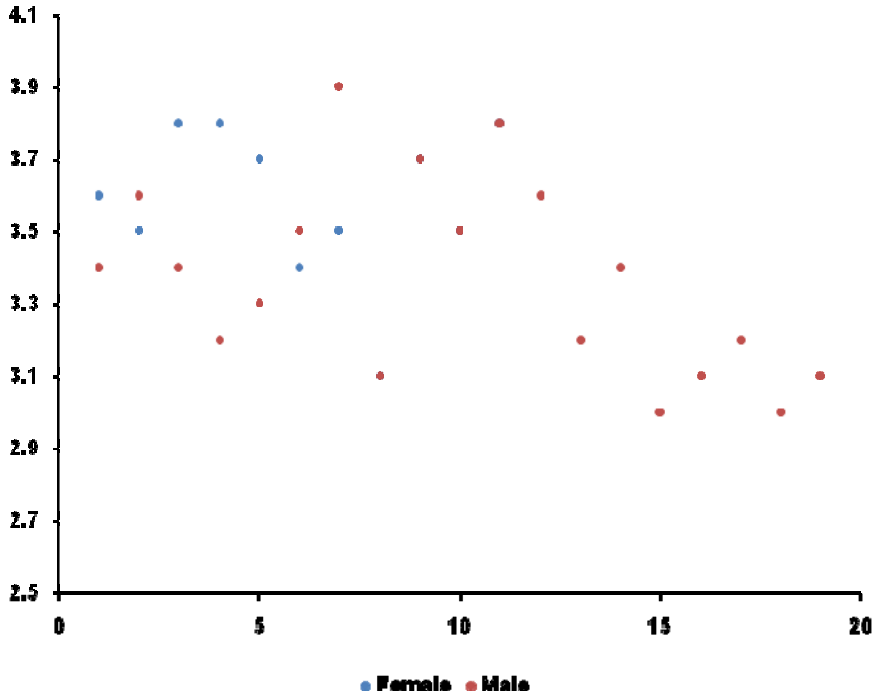
On analyzing the significance of gender on the eight parameters in the PATIENT group, the results were as follows.

Table 3
Chiari Group Statistics

	Group	N	Mean	Std. Deviation	Std. Error Mean
VOLUME	Female	11	240.327	19.406	5.851
	Male	19	239.264	17.028	3.906
BASIOCCIPUT	Female	11	2.645	.375	.113
	Male	19	2.711	.283	.065
FORAMEN MAGNUM	Female	11	3.582	.214	.064
	Male	19	3.368	.269	.062
SUPRAOCCIPUT	Female	11	4.093	.135	.041
	Male	19	4.093	.129	.029
TENTORIAL ANGLE	Female	11	85.118	.916	.276
	Male	19	85.100	1.235	.283
BRAINSTEM LENGTH	Female	11	4.889	.174	.053
	Male	19	4.956	.125	.029
CEREBELLAR LENGTH	Female	11	4.928	.137	.041
	Male	19	4.892	.119	.027
HEIGHT	Female	11	6.491	.263	.079
	Male	19	6.632	.316	.073

Table 4
Chiari Group Independent Samples Test

		t-test for Equality of Means				
					95% Confidence Interval of the Difference	
		t	df	Sig. (2-tailed)	Lower	Upper
VOLUME	Equal variances assumed	.157	28.000	.877	-12.839	14.966
	Equal variances not assumed	.151	18.824	.881	-13.671	15.798
BASIOCCIPUT	Equal variances assumed	-.539	28.000	.594	-.312	.182
	Equal variances not assumed	-.499	16.654	.624	-.341	.210
FORAMEN MAGNUM	Equal variances assumed	2.248	28.000	.033	.019	.408
	Equal variances not assumed	2.393	25.047	.025	.030	.397
SUPRAOCCIPUT	Equal variances assumed	.002	28.000	.998	-.102	.102
	Equal variances not assumed	.002	20.148	.999	-.105	.105
TENTORIAL ANGLE	Equal variances assumed	.042	28.000	.966	-.860	.896
	Equal variances not assumed	.046	26.074	.964	-.795	.832
BRAINSTEM LENGTH	Equal variances assumed	-1.217	28.000	.234	-.179	.046
	Equal variances not assumed	-1.113	16.034	.282	-.194	.060
CEREBELLAR LENGTH	Equal variances assumed	.767	28.000	.450	-.061	.134
	Equal variances not assumed	.738	18.685	.470	-.067	.141
HEIGHT	Equal variances assumed	-1.245	28.000	.223	-.372	.091
	Equal variances not assumed	-1.310	24.328	.202	-.362	.081



Scatter chart - FORAMENMAGNUM - Chiari group

There is statistically significant difference in the AP diameter of the Foramen magnum between males (mean – 3.36 cm) and females (mean – 3.58 cm) in the patient group (P - value – 0.033).

There was no significant difference in the other measured parameters between males and females in the patient group.

The tables below show the mean values and statistical analysis of the eight parameters between females in the Chiari and control group

Table. 5

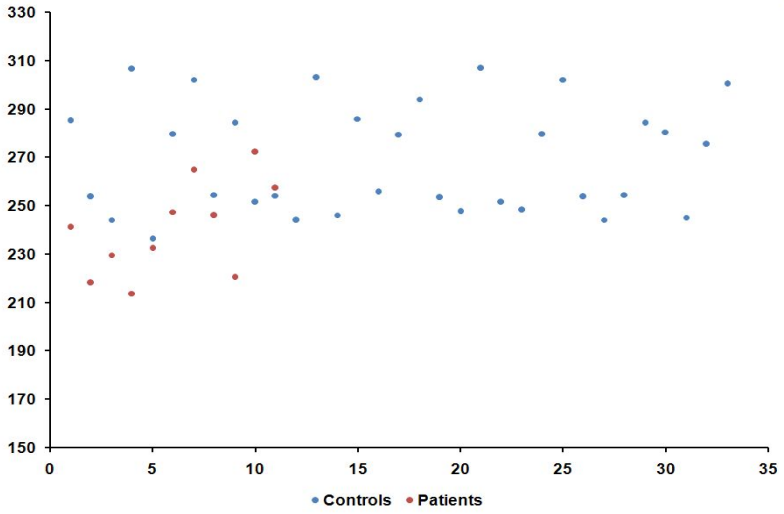
	Group Statistics				
	Group	**N**	**Mean**	**Std. Deviation**	**Std. Error Mean**
VOLUME	Control	33	269.40	22.63	3.94
	Chiari	11	240.33	19.41	5.85
BASIOCCIPUT	Control	33	2.75	0.33	0.06
	Chiari	11	2.65	0.38	0.11
FORAMEN MAGNUM	Control	33	3.42	0.36	0.06
	Chiari	11	3.58	0.21	0.06
SUPRAOCCIPUT	Control	33	4.08	0.12	0.02
	Chiari	11	4.09	0.14	0.04
TENTORIAL ANGLE	Control	33	84.31	0.86	0.15
	Chiari	11	85.12	0.92	0.28
BRAINSTEM LENGTH	Control	33	4.88	0.16	0.03
	Chiari	11	4.89	0.17	0.05
CEREBELLAR LENGTH	Control	33	4.92	0.14	0.02
	Chiari	11	4.93	0.14	0.04
HEIGHT	Control	33	7.12	0.37	0.06
	Chiari	11	6.49	0.26	0.08

Table 6

Female - Independent Samples Test								
		Levene's Test for Equality of Variances		t-test for Equality of Means				
							95% Confidence Interval of the Difference	
		F	Sig.	t	df	Sig. (2-tailed)	Lower	Upper
VOLUME	Equal variances assumed	2.372	0.131	3.812	42.000	0.000	13.683	44.464
	Equal variances not assumed			4.122	19.845	0.001	14.353	43.795
BASIOCCIPUT	Equal variances assumed	0.265	0.609	0.886	42.000	0.380	-0.135	0.348
	Equal variances not assumed			0.834	15.613	0.417	-0.164	0.376
FORAMEN MAGNUM	Equal variances assumed	1.314	0.258	-1.384	42.000	0.174	-0.397	0.074
	Equal variances not assumed			-1.785	30.033	0.084	-0.346	0.023
SUPRAOCCIPUT	Equal variances assumed	0.316	0.577	-0.407	42.000	0.686	-0.105	0.070
	Equal variances not assumed			-0.384	15.644	0.706	-0.115	0.080
TENTORIAL ANGLE	Equal variances assumed	0.280	0.599	-2.654	42.000	0.011	-1.419	-0.193
	Equal variances not assumed			-2.566	16.270	0.021	-1.471	-0.141
BRAINSTEM LENGTH	Equal variances assumed	0.411	0.525	-0.119	42.000	0.906	-0.120	0.106
	Equal variances not assumed			-0.113	15.735	0.912	-0.132	0.119
CEREBELLAR LENGTH	Equal variances assumed	0.007	0.934	-0.205	42.000	0.838	-0.105	0.086
	Equal variances not assumed			-0.204	16.945	0.841	-0.110	0.091
HEIGHT	Equal variances assumed	0.026	0.873	5.229	42.000	0.000	0.389	0.877
	Equal variances not assumed			6.201	24.324	0.000	0.422	0.844

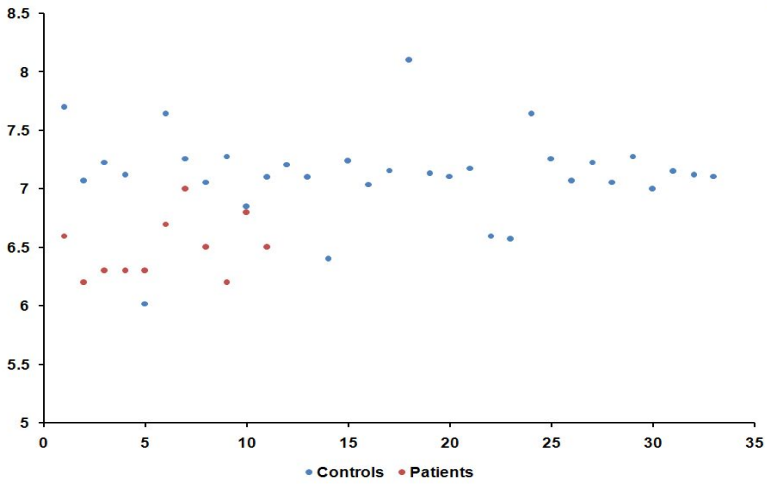
The scatter charts for the statistically significant parameters is shown below

Scatter chart VOLUME (Females)



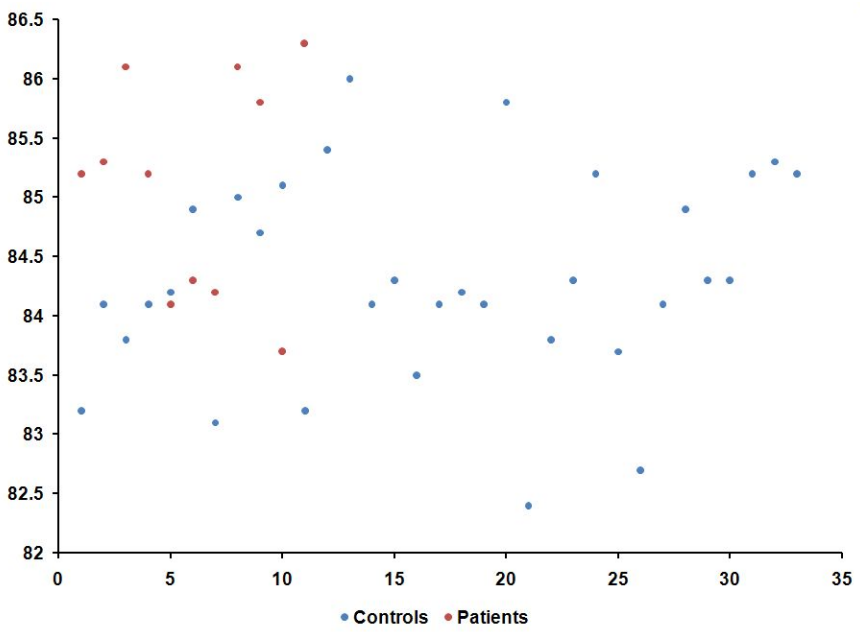
For volume there is statistically significant difference between Controls(mean – 269.4 cc) and Chiari group(mean – 240.3 cc) (P-Value < 0.05).

Scatter chart HEIGHT (Females)



For height there is statistically significant difference between Controls(mean – 7.12 cm) and Chiari group(mean -6.49 cm) (P-Value < 0.05)

Scatter chart TENTORIAL ANGLE (Females)



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For tentorial angle there is statistically significant difference betwen Controls(mean – 84.3^0) and Chiari group(mean - 85.1^0) (P-Value - 0.011).

There was no statistical significance in the values of other five parameters.

The tables below shows a similar analysis between males in the Chiari and control group.

Table 7

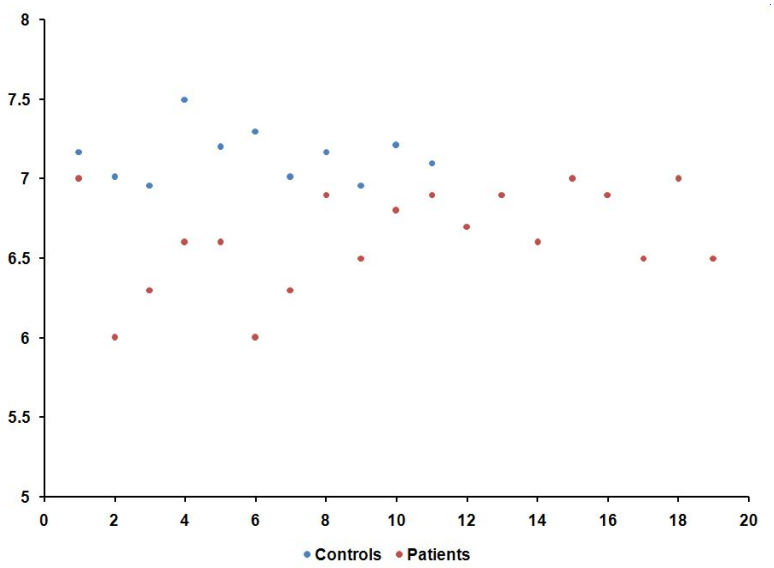
	Group Statistics				
	GROUP	**N**	**Mean**	**Std. Deviation**	**Std. Error Mean**
VOLUME	Control	11	287.93	21.00	6.33
	Chiari	19	239.26	17.03	3.91
BASIOCCIPUT	Control	11	2.68	0.57	0.17
	Chiari	19	2.71	0.28	0.06
FORAMEN MAGNUM	Control	11	3.38	0.20	0.06
	Chiari	19	3.37	0.27	0.06
SUPRAOCCIPUT	Control	11	4.11	0.11	0.03
	Chiari	19	4.09	0.13	0.03
TENTORIAL ANGLE	Control	11	84.35	0.85	0.26
	Chiari	19	85.10	1.24	0.28
BRAINSTEM LENGTH	Control	11	4.83	0.15	0.05
	Chiari	19	4.96	0.12	0.03
CEREBELLAR LENGTH	Control	11	4.99	0.14	0.04
	Chiari	19	4.89	0.12	0.03
HEIGHT	Control	11	7.14	0.16	0.05
	Chiari	19	6.63	0.32	0.07

Table 8 Male - Independent Samples Test

		Levene's Test for Equality of Variances		t-test for Equality of Means				
							95% Confidence Interval of the Difference	
		F	Sig.	t	df	Sig. (2-tailed)	Lower	Upper
VOLUME	Equal variances assumed	1.417	0.244	6.927	28.000	0.000	34.274	63.057
	Equal variances not assumed			6.541	17.642	0.000	33.012	64.319
BASIOCCIPUT	Equal variances assumed	10.986	0.003	-0.168	28.000	0.868	-0.343	0.291
	Equal variances not assumed			-0.142	12.929	0.889	-0.422	0.370
FORAMEN MAGNUM	Equal variances assumed	1.495	0.232	0.114	28.000	0.910	-0.181	0.202
	Equal variances not assumed			0.123	25.980	0.903	-0.167	0.188
SUPRAOCCIPUT	Equal variances assumed	0.675	0.418	0.375	28.000	0.710	-0.077	0.112
	Equal variances not assumed			0.392	23.811	0.699	-0.074	0.109
TENTORIAL ANGLE	Equal variances assumed	2.645	0.115	-1.768	28.000	0.088	-1.609	0.118
	Equal variances not assumed			-1.952	27.000	0.061	-1.529	0.038
BRAINSTEM LENGTH	Equal variances assumed	0.874	0.358	-2.473	28.000	0.020	-0.233	-0.022
	Equal variances not assumed			-2.335	17.622	0.032	-0.243	-0.013
CEREBELLAR LENGTH	Equal variances assumed	0.308	0.583	2.095	28.000	0.045	0.002	0.198
	Equal variances not assumed			2.014	18.619	0.059	-0.004	0.205
HEIGHT	Equal variances assumed	4.944	0.034	4.984	28.000	0.000	0.302	0.724
	Equal variances not assumed			5.854	27.777	0.000	0.333	0.693

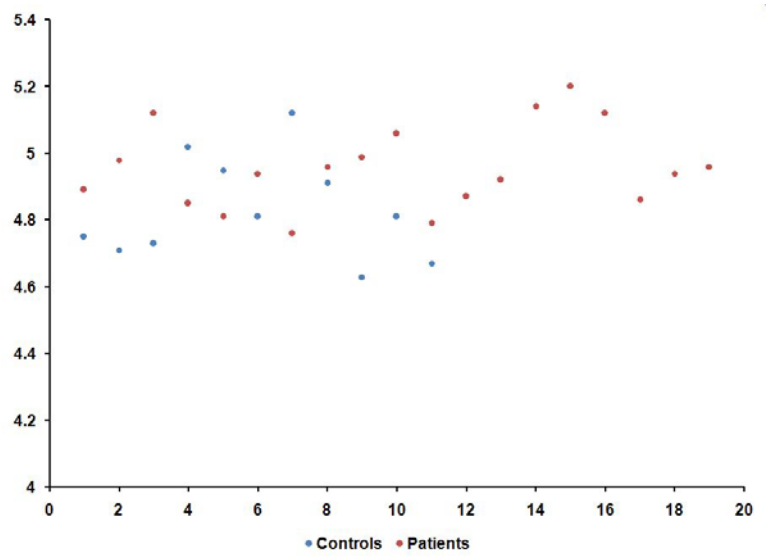
The Scatter charts for the Statistically significant parameters is shown below

Scatter chart HEIGHT (Males)



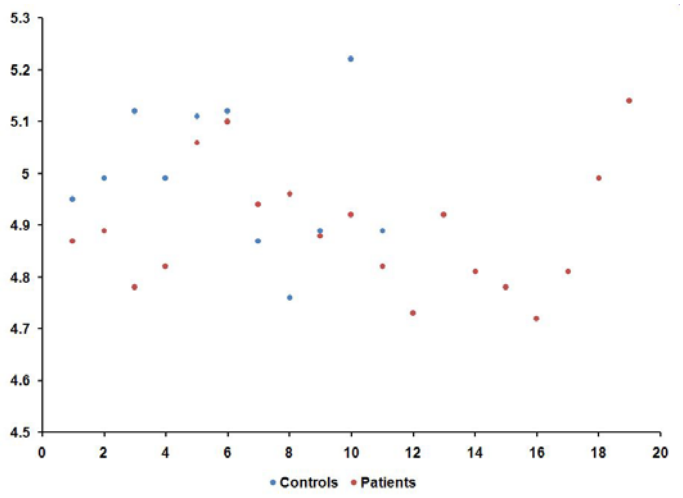
For height there is statistically significant difference between controls(mean – 7.14 cm) and Chiari group (mean -6.63 cm)(P-Value - 0.000)

Scatter chart CEREBELLAR LENGTH (Males)



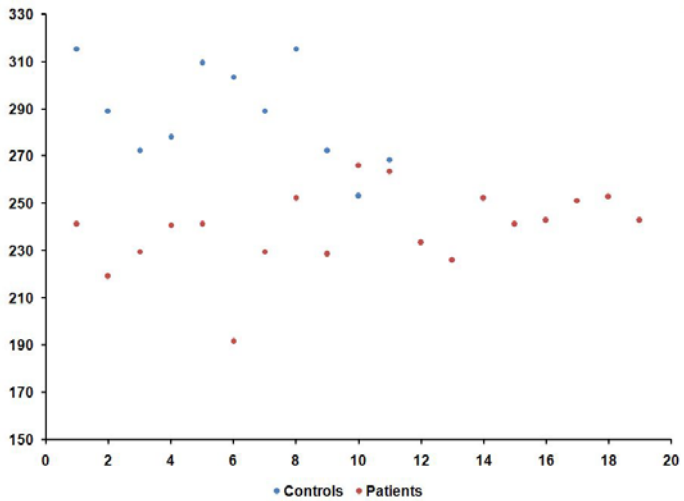
For cerebellar length there is statistically significant difference between controls(mean -4.99 cm) and Chiari group (mean -4.89 cm) (P-value - 0.045).

Scatter chart BRAINSTEM LENGTH (Males)



For brainstem length there is statistically significant difference between Controls(mean – 4.83 cm) and Chiari group(mean -4.96cm) (P-value - 0.020).

Scatter Diagram VOLUME (Males)



For volume there is statistically significant difference between Controls(mean – 287.9 cc) and Chiari group(mean -239.2 cc) (P-Value - 0.000).

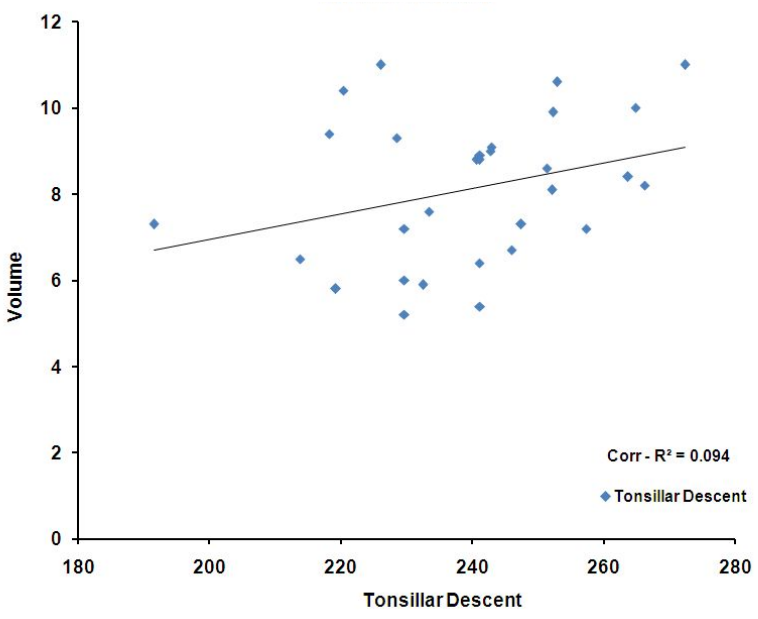
There was no statistical significance in the values of other parameters.

In Chiari patients the mean value of tonsillar descent was 8.1 mm. In controls the tonsil was above the McRae's line.

Table. 9

		Volume	**Tonsillar Descent**
Volume	Pearson Correlation	1	.308
	Sig. (2 – tailed)	.	.098
	N	30	30
Tonsillar Descent	Pearson Correlation	.308	1
	Sig. (2-tailed)	.098	.
	N	30	30

Scatter chart VOLUME VS TONSILLAR DESCENT



On analysing the relationship between the volume of PCF and the extent of tonsilllar descent there was no relationship between the two parameters.

DISCUSSION

Various theories have been postulated to explain the pathogenesis of Chiari 1 malformation.Most accepted theory is a primarily small posterior cranial fossa.To establish this in the Indian population , an attempt has been made and various parameters have been analysed.

To estimate overcrowding in the posterior cranial fossa of patients with Chiari 1 malformation, we calculated the volume and other linear measurements of the bony and neural structures of the posterior cranial fossa.

The mean volume of the PCF in this study was 274 ± 23 cc, in the control group. In the patient group the mean volume was 239 ±17 cc. The volume of the Chiari group was significantly smaller than the control group even on further gender analysis. Three morphometric studies in the past have suggested a significantly smaller volume of the posterior cranial fossa in adult patients with Chiari malformation. Previous volumetric reports on the assessment of cranioencephalic disproportion have been discordant whatever the methodology of PCF volume calculation.

Using the Cavalieri method, Milhorat et al.[31] have confirmed the disproportion, showing a significant reduction of PCF volume in patients with CMI but no differences in brain volumes compared with controls.

Nishikawa et al.[33] have found no significant difference in the PCF volume between CM-I and control groups, but they did determine that the volume ratio between PCF brain and cranial volumes was significantly increased in CM-I. In Nishikawas study the mean PCF volume as measured by CT scanning was 186 cubic centimeters in the Chiari group (188 cubic centimeters in patients without vs. 184 cubic centimeters in those with basilar invagination) and 193 cubic centimeters in the control group. With a different methodology, Vega et al.[45] have demonstrated a significant reduction in PCF volumes only in males.

Morphometric study by Menezes et al.[21] has revealed a mean volume of 259 ± 41 cc (range 187 - 323cc)in the patient group whereas in the control group the average volume was 355 ± 46 cc (range 274 - 415cc). Despite the use of advanced image segmentation techniques for PCF volume calculation, the results remain controversial. The average height of the PCF was shorter in the Chiari group(6.58 cm) compared to the Control group(7.13 cm).This is in concordance with a previous morphometric study by Menezes et al.[21] in which the mean value of the height of the PCF was 5.8 cm in the patient group and 7.5 cm in the Control group . The present study clearly showed overcrowding of the posterior cranial fossa, in Chiari 1 malformation, by direct measurement of the volume of the posterior cranial fossa. Downward herniation of the normally developing hindbrain occurs due to the overcrowding in the posterior cranial fossa

O'Rahilly and colleagues[34] have proved that the basioccipital and exoccipital portions of the occipital bone up to the top of the jugular tubercle are derived from the occipital somites in the human fetus The present study took into consideration the embryological development of the human occipital enchondrium, and each part of the base of the skull in adult patients with Chiari 1 malformation was compared to that of normal controls.

The mean length of the basiocciput was 2.69 cm in the Chiari group and 2.73 cm in the control group , in this study. Although the mean length of the basiocciput was less in the Chiari group, it was statistically insignificant .The mean length of the basiocciput in the CM-I and control groups were 1.94 cm and 2.57 cm , respectively in Noudels study.[38]

All the previous authors have reported a decreased length of the basiocciput/clivus in CM-I group except Nishikawa et al[33] study in which a shortened basiocciput was found only in patients who had CM-I and basilar invagination.

The length of the supraocciput was 4.08 cm in the control group and 4.09 cm in the patient group and was statistically insignificant on a gross comparative analysis and further gender analysis.These values are consistent with the ones reported , in Noudels study[38].The statistical insignificance is in concordance with the three morphometric studies by Aydin et al[4],Sekula et al[40] and Noudel et al[38].On the contrary Nishikawa et al[33] and Milhorat et al[31] have demonstrated a significant shortening of the supraocciput in CM-I patients.

The present study also showed that the cerebellar tentorium in the Chiari group was significantly steeper than in the control group. The mean angle of the cerebellar tentorium was 85.11^0 in the Chiari group and 84.32^0 in the control group and the tentorium was significantly steeper on further gender analysis.

In Milhorat's study[31] the mean angle of the Tentorium in the Chiari and Control groups were 90^0 and 82.5^0 and were statistically significant.In Noudel's study[38] the mean tentorial angle was 85.3^0 and 84.6^0 in the CM-I and Control groups respectively and were statistically insignificant. Overcrowding in the posterior cranial fossa may induce a consistent upward shifting of its contents, leading to a significantly steeper tentorium as well as a downward shifting of the hindbrain. Despite the lack of significant abnormality of any other osseous structure of the PCF in our study, it has been shown that caudal displacement of the hindbrain can shift upward the cerebellar tentorium.

The mean anteroposterior diameter of the foramen magnum was 3.45 cm in the Chiari group and 3.41 cm in the control group and were statistically insignificant .These values compare well with the AP diameter of foramenmagnum in Noudel's study[38] .Aydin et al[4] have documented a AP diameter of 3.17 cm and 2.52 cm in Chiari group and Control group respectively. These were statistically significant. On further gender analysis the AP diameter of the foramen magnum was significantly longer in females compared to males in the patient group. It has been shown that caudal displacement of the hindbrain can enlarge the foramen magnum .

In the present study the mean axial lengths of the brainstem and cerebellum were 4.87 cm and 4.94 cm in the control group. In the Chiari group the values were 4.93 cm and 4.91 cm respectively. These values compare well with the values documented by previous studies. In the present study , there was no significant difference in the long axial lengths of the hindbrain (pons, medulla oblongata, and cerebellum) in females and both groups on the whole.

On a comparative analysis between males in the Chiari and patient group the mean axial length of the brainstem was 4.96 cm and 4.83 cm respectively. The cerebellar length was 4.89 cm and 4.99 cm in the Chiari and patient groups respectively. These were statistically significant in this subset. None of the published literature have documented a significant difference in the dimensions of the neural structures . In the Indian population it has been observed that brainstem and cerebellar length is altered in the male subset of patients. The relevance of this finding needs to be confirmed by a larger study sample.

In Chiari patients the mean value of tonsillar descent was 8.1 mm . The mean value of tonsillar descent was 9.8 mm in Milhorat's study[31]. In the control group the tonsil was above the McRae's line. This is in discordance with Milhorat's study[31] in which the mean tonsillar descent was 2.1 mm in the Control group. The degree of tonsillar herniation does not seem to be representative of the cranioencephalic disproportion. In

Nishikawa's study[33] no significant relationship was found between the volume of the herniated brain below the foramen magnum and the PCF volume or between the volume of the herniated brain below the foramen magnum and the volume ratio.

Our data failed to show any significant correlation between the extent of tonsillar ectopia and any other measured parameters of the PCF, as has Vega's[45] and Noudels[38] studies. However, Schady[39] et al have found an inverse relationship between the size of the PCF and the degree of cerebellar herniation, whereas Stovner et al.[42] have shown a strong positive correlation between the two parameters.

Furthermore, our results confirm that the most consistent MR imaging finding is compression of the retrocerebellar CSF spaces of the cisterna magna by the herniated tonsils, which provides substantial evidence of overcrowding. This finding has been documented by Milhorat et al[31] , Aydin et al[4] and Noudel et al[38] .

The table below shows a comparison between our study and the published literature.

Table 10 : Literature review of morphometric studies of the posterior cranial fossa*

Authors & Year	Radiographic Studies	Clivus or Basiocciput	SO	PCF Volume	Tentorial Angle	Neural Structures	Cisterna Magna
Schady et al., 1987	Radio + CT	↓ (NS for women)	-	__	__	__	__
Vega et al., 1990	Radio + CT	↓ (for CMI + BI)	_	↓ (NS for women)	__	__	__
Nishikawa et al., 1997	MRI	↓ (for CMI + BI)	↓	↓ NS	↑	Normal	__
Mihorat et al., 1999	MRI	↓	↓	↓	↑	Normal	absent
Karagoz et al., 2002	MRI	↓	__	__	↑	__	__
Aydin et al., 2005	MRI	↓	↓ NS	__	__	__	Absent
Sekula et al., 2005	MRI	↓	↓ NS	__	↑	Normal	Absent
Noudel et al	MRI	↓	__	__	↑ NS	Normal	Absent
Present study	MRI	↓ NS	No difference	↓	↑	Normal (except in males)	Absent

BI – Basilar invagination;NS – Not significant

In the present study, MRI findings of reduced height of the PCF and increased slope of the tentorium,are consistent with a defect of the para-axial mesoderm.

Milhorat et al. and Nishikawa et al.[31,32,33] have postulated that the fundamental pathogenic entity in CMI is most likely underdevelopment of the

para-axial mesoderm resulting in posterior fossa hypoplasia with CSF flow abnormalities.

Much evidence supports the theory that CM-I is primarily a mesodermal developmental abnormality[5,10,29] . Marin-Padilla et al. first observed that the posterior cranial fossa is hypoplastic in Chiari I malformation[26,27] . Multiple morphometric studies have since implicated overcrowding of hindbrain by an underdeveloped posterior cranial fossa in the development of CM-I. Overcrowding of the hindbrain and resulting displacement of CSF likely contributes to the array of symptoms seen in CM-I.

Since the volume of PCF showed a significant correlation it indicates that decompression of the posterior cranial fossa may relieve overcrowding of the posterior cranial fossa. It can be inferred that overcrowding of the posterior cranial fossa induces remodeling of neural structures as the cerebellar tentorium shifts upward and the cerebellar tonsils herniate to accommodate the growing brain, rather than remodeling the cranium.

It could also be postulated that this altered geometry of the posterior fossa that initiates the downward migration is subsequently perpetuated by CSF movement and pulsation.

Consequently,most clinical symptoms result from displacement of newly formed CSF from the subarachnoid spaces of the PCF into available spaces within the supratentorial and spinal compartments. Current evidence[38]

suggests that hindbrain-related syringomyelia, observed in 60% of our patients, is also a complication resulting from obstructed CSF flow between the cranial and spinal compartments.

The considerable delay in the occurrence of neurological symptoms could be explained by the relatively late, mainly postnatal, growth spurt of the cerebellum within a small and inadequate PCF.

Thus, when paraxial mesodermal insufficiency is regarded as the pathogenesis of Chiari malformation, adult-type Chiari malformation can be considered a mild form and the pediatric type a severe form.

Badie et al. have demonstrated that patients with smaller posterior fossa volumes presented at a younger age and had a better response to surgery. Cardiac-gated phase-contrast cine MR can be a valuable tool in identifying patients who are less likely to respond to suboccipital decompression for CMI.

Morphometric and volumetric studies are useful tools in increasing the understanding of the pathophysiological conditions at play in the development of CTH. Morphometric parameters of the posterior cranial fossa could influence the natural history of patients with CMI as well as their prognosis after surgical treatment and could reduce the risk of postoperative complications such as cerebellar ptosis.

Further studies are needed to identify preoperatively the steps of the surgical treatment and the bone resection needed for decompression of the posterior fossa in the individual. This is the first morphometric study of the PCF in the Indian adult population. A more extensive study, using repeatable measures with a larger number of patients, is necessary to confirm the results of this study and evaluate their further application.

However this is only a preliminary study with a relatively small sample size and a larger study with more number of patients will throw more light into the pathogenesis of CM-I.This study has been done only in adult population. A separate study is needed for pediatric population with CM-I.

CONCLUSION

Morphometric analysis of posterior cranial fossa in healthy adult population and adult patients with Chiari 1 malformation in the Indian population has led us to the following conclusions.

1. The volume of posterior cranial fossa in healthy adult population is 251cc – 297cc and the volume of posterior cranial fossa in adult patients with Chiari 1 malformation is 222cc – 256cc. and the volume of posterior cranial fossa in adult patients with Chiari 1 malformation is significantly smaller than the healthy adult population.The volume of posterior cranial fossa in healthy Indian adult population is smaller than the western population.

2. The height of the posterior cranial fossa in adult patients with Chiari 1 malformation is significantly smaller than the healthy adult population.

3. The tentorium in adult patients with Chiari 1 malformation is steeper than the healthy adult population.

4. Though the mean length of the basiocciput is smaller in adult patients with Chiari 1 malformation, it is not statistically significant.

5. There is no significant difference in the sagittal diameter of the foramen magnum between Chiari patients and healthy adult population.

This study reestablishes the fact that CM-I is a disorder of the mesoderm and the fundamental problem in CM-I is a volumetrically small posterior cranial fossa which results in varying degrees of hindbrain overcrowding.

APPENDIX – I
PROFORMA

Name Age Sex I.P. No M.I.N No Unit

Contact Address / No

D.O.A.

D.O.S

D.O.D

CLINICAL DATA

SYMPTOMS & DURATION

NEUROLOGICAL DEFICIT

RADIOLOGICAL PARAMETERS

x - *maximum width of bony posterior cranial fossa*

y - *distance from posterior clinoid process to torcula heterophili*

z - *Height of Posterior Cranial fossa - from basion to peak of tentorium cerebelli*

PCF Volume - 4/3 π x (x/2 x y/2 x z/2)

*Length of basiocciput (**a**)*

*Length of supraocciput (**b**)*

*Tentorial angle (**c**)*

*Brainstem length (**d**)*

*Cerebellar length (**e**)*

*Length of tonsillar descent (**f**)*

Cisterna magna

APPENDIX - II **Excel Chart (Control Group)**

AGE (yrs)	SEX	WIDTH (cm)	HEIGHT (cm)	SAGGITAL AP (cm)	VOLUME (cc)	BASIOCCIPUT (cm)	FORAMEN MAGNUM (cm)	SUPRAOCCIPUT (cm)	TENTORIAL ANGLE	BRAINSTEM LENGTH (cm)	CEREBELLAR LENGTH (cm)
21	F	9.26	7.7	8	285.2	2.5	2	4	83.2	4.87	5.12
38	F	9.17	7.07	7.83	253.81	2.27	3.07	4.1	84.1	4.92	4.92
25	M	10.14	7.17	8.68	315.53	2.23	3.15	3.97	84.2	4.75	4.95
34	F	8.96	7.23	7.54	244.23	2.96	3.24	4.06	83.8	4.93	4.72
27	F	10.17	7.12	8.47	306.65	2.6	3.34	4.1	84.1	5.1	4.83
57	F	10.43	6.02	7.53	236.39	2.39	3.97	4.14	84.2	4.96	4.81
43	F	9.9	7.64	7.4	279.85	2.57	3.4	4.01	84.9	4.95	4.91
62	M	9.85	7.01	8.38	289.31	2.53	3.49	4.09	83.7	4.71	4.99
32	F	9.68	7.26	8.6	302.19	2.18	3.24	4.12	83.1	4.84	5.12
30	M	9.83	6.96	7.96	272.29	3.63	3.24	4.11	85.1	4.73	5.12
23	F	9.48	7.06	7.61	254.66	3.37	3.49	4.09	85	4.87	4.91
30	F	9.56	7.28	8.17	284.3	3.24	3.23	3.99	84.7	4.92	4.99
46	F	9.66	6.85	7.6	251.44	3.23	3.32	4.17	85.1	4.96	4.96
37	M	9.34	7.5	7.94	278.09	3.14	3.36	4.2	83.1	5.02	4.99
40	F	9.15	7.1	7.83	254.33	2.7	3.7	4.31	83.2	5.1	5.21
23	M	10	7.2	8.6	309.6	2.18	3.67	4.17	83.6	4.95	5.11
36	F	8.98	7.21	7.54	244	2.78	3.6	4.21	85.4	4.87	4.89
30	F	10.04	7.1	8.5	302.95	2.62	3.35	4.24	86	4.83	4.88
50	F	10.23	6.4	7.52	246.17	3.04	3.13	3.96	84.1	4.99	4.99
45	F	9.66	7.24	8.18	286.04	3	3.78	3.98	84.3	5.02	4.72
32	F	9.47	7.04	7.67	255.67	3	3.64	4.04	83.5	5.2	4.81
40	F	10	7.16	7.81	279.59	2.7	3.51	4.15	84.1	4.91	4.83
34	F	9.24	8.1	7.86	294.13	2.6	3.4	4.16	84.2	4.82	4.91
40	F	9.15	7.13	7.77	253.45	2.75	3.8	4.19	84.1	4.73	4.99
23	M	9.9	7.3	8.4	303.53	2.25	3.74	4.23	84.2	4.81	5.12
36	F	9.24	7.11	7.54	247.67	2.7	3.72	3.89	85.8	4.68	5.11
30	F	9.94	7.18	8.6	306.88	2.52	3.25	3.85	82.4	4.53	5.12
50	F	10	6.6	7.62	251.46	3	3.17	3.98	83.8	4.73	4.9
57	F	10.2	6.57	7.41	248.28	2.39	3.97	4	84.3	4.82	4.72
43	F	9.9	7.64	7.4	279.85	2.57	3.4	4.04	85.2	4.91	4.91
62	M	9.85	7.01	8.38	289.31	2.53	3.49	4.02	84.1	5.12	4.87
32	F	9.68	7.26	8.6	302.19	2.18	3.24	4.32	83.7	5.23	4.97
38	F	9.17	7.07	7.83	253.81	2.27	3.07	3.8	82.7	4.76	4.99
25	M	10.14	7.17	8.68	315.53	2.23	3.15	3.89	84.3	4.91	4.76
34	F	8.96	7.23	7.54	244.22	2.96	3.24	3.97	84.1	4.52	4.63
30	M	9.83	6.96	7.96	272.29	3.63	3.24	4.2	84.2	4.63	4.89
23	F	9.48	7.06	7.61	254.66	3.37	3.49	3.99	84.9	4.75	4.88
30	F	9.56	7.28	8.17	284.3	3.24	3.23	4.1	84.3	4.76	4.9

28	F	10.02	7	8	280.56	2.56	3.9	4.21	84.3	4.87	5.12
32	M	9.24	7.21	7.6	253.15	2.2	3.24	4.12	85.3	4.81	5.22
26	F	9.12	7.15	7.52	245.18	2.96	3.84	4.14	85.2	4.98	4.94
41	M	9.7	7.1	7.8	268.59	2.98	3.4	4.21	86.1	4.67	4.89
35	F	9.9	7.12	7.82	275.6	2.72	3.54	4.11	85.3	4.87	4.82
31	F	9.52	7.11	8.88	300.53	2.86	3.6	4.06	85.2	4.92	4.78

APPENDIX - II Excel Chart (Chiari Group)

AGE (yrs)	SEX	WIDTH (cm)	HEIGHT (cm)	SAGGITAL AP (cm)	VOLUME (cc)	BASIOCCIPUT (cm)	FORAMEN MAGNUM (cm)	SUPRAOCCIPUT (cm)	TENTORIAL ANGLE	BRAINSTEM LENGTH (cm)	CEREBELLAR LENGTH (cm)	Tonsillar Descent (cm)	Cisterna magna
45	M	8.4	7	8.2	241.08	2.1	3.4	4.24	83.2	4.89	4.87	6.4	-
18	M	8.7	6	8.4	219.24	2.5	3.6	4.21	86.1	4.98	4.89	5.8	-
55	M	9	6.3	8.1	229.63	2.6	3.4	3.94	87.3	5.12	4.78	7.2	-
36	F	8.7	6.6	8.4	241.16	2.7	3.6	3.87	85.2	5.21	4.97	5.4	-
20	F	8.8	6.2	8	218.24	2.1	3.5	3.97	85.3	4.91	4.91	9.4	-
32	M	9	6.6	8.1	240.57	2.5	3.2	4.23	84.3	4.85	4.82	8.8	-
27	F	9	6.3	8.1	229.63	2	3.8	4.21	86.1	4.76	4.99	6	-
23	M	8.7	6.6	8.4	241.16	2.6	3.3	4.11	86.3	4.81	5.06	8.9	-
21	M	8.4	6	7.6	191.52	2.8	3.5	4.2	87.1	4.94	5.1	7.3	-
21	F	8.7	6.3	7.8	213.75	3	3.8	3.99	85.2	4.81	4.87	6.5	-
28	M	9	6.3	8.1	229.63	2.4	3.9	3.91	84.2	4.76	4.94	5.2	-
26	F	9	6.3	8.2	232.47	2.9	3.7	4.12	84.1	4.62	4.78	5.9	-
36	F	8.9	6.7	8.3	247.46	2.7	3.4	4.15	84.3	4.86	5.12	7.3	-
32	M	8.7	6.9	8.4	252.12	3	3.1	4.05	85.2	4.96	4.96	8.1	-
30	M	8.9	6.5	7.9	228.5	2.9	3.7	4.21	84.2	4.99	4.88	9.3	-
24	M	9	6.8	8.7	266.22	2.7	3.5	3.82	84.1	5.06	4.92	8.2	-
36	F	8.8	7	8.6	264.88	3.1	3.5	3.95	84.2	5.12	4.91	10	-
26	M	9.1	6.9	8.4	263.71	3.3	3.8	4	84.1	4.79	4.82	8.4	-
30	M	8.6	6.7	8.1	233.36	2.8	3.6	3.91	84.7	4.87	4.73	7.6	-
23	F	8.8	6.5	8.6	245.96	3	3.1	4.3	86.1	4.97	4.86	6.7	-
29	M	8.4	6.9	7.8	226.04	2.4	3.2	4.11	86.9	4.92	4.92	11	-
36	F	9	6.2	7.9	220.41	2.5	3.7	4.25	85.8	4.99	4.78	10.4	-
36	M	8.4	6.6	9.1	252.25	2.8	3.4	4.18	85.3	5.14	4.81	9.9	-
44	M	8.4	7	8.2	241.08	2.5	3	4.11	84.9	5.2	4.78	8.8	-
30	M	8.8	6.9	8	242.88	2.9	3.1	4.21	85.3	5.12	4.72	9.1	-
28	M	8.5	6.5	9.1	251.38	3.1	3.2	4.01	86	4.86	4.81	8.6	-
20	F	8.9	6.8	9	272.34	2.8	3.5	4.13	83.7	4.81	4.81	11	-
31	M	8.5	7	8.5	252.87	2.7	3	4.21	83.2	4.94	4.99	10.6	-
40	M	9	6.5	8.3	242.77	2.9	3.1	4.1	84.5	4.96	5.14	9	-
22	F	8.7	6.5	9.1	257.3	2.3	3.8	4.08	86.3	4.72	5.21	7.2	