CLINICAL AND RADIOLOGICAL OUTCOMES FOLLOWING FORAMEN MAGNUM DECOMPRESSION IN PATIENTS WITH CHIARI MALFORMATION

Submitted for PDF Skullbase Neurosurgery

By

Dr. JAYPALSINH GOHIL
JAN 2020 - DEC 2020

DEPARTMENT OF NEUROSURGERY

SREE CHITRA TIRUNAL INSTITUTE FOR MEDICAL SCIENCES
& TECHNOLOGY

THIRUVANANTHAPURAM – 695011

CLINICAL AND RADIOLOGICAL OUTCOMES FOLLOWING FORAMEN MAGNUM DECOMPRESSION IN PATIENTS WITH CHIARI MALFORMATION

Submitted by : Dr. Jaypalsinh Gohil

Programme : PDF skullbase Neurosurgery

Month & year of submission : Dec- 2020

DECLARATION

This thesis titled — CLINICAL AND RADIOLOGICAL OUTCOMES FOLLOWING FORAMEN MAGNUM DECOMPRESSION IN PATIENTS WITH CHIARI MALFORMATION is a consolidated report based on a bonafide study of the period from January 2020 to December 2020, done by me under the Department of Neurosurgery, Sree Chitra Tirunal Institute for Medical Sciences & Technology, Thiruvananthapuram.

This thesis is submitted to SCTIMST in fulfillment of rules and regulations of the PDF skullbase Neurosurgery course.

J.A. Gohil

Dr. Jaypalsinh Gohil,

Department of Neurosurgery,

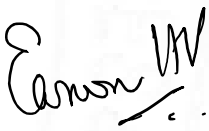
SCTIMST, Thiruvananthapuram.

CERTIFICATE

This is to certify that the thesis entitled — **CLINICAL AND RADIOLOGICAL OUTCOMES FOLLOWING FORAMEN MAGNUM DECOMPRESSION IN PATIENTS WITH CHIARI MALFORMATION"** is a bonafide work of Dr. Jaypalsinh Gohil and was conducted in the Department of Neurosurgery, Sree Chitra Tirunal Institute for Medical Sciences & Technology, Thiruvananthapuram (SCTIMST), under my guidance and supervision.

Dr. Ganesh Divakar,
Assistant Professor,
Department of Neurosurgery,
SCTIMST,
Thiruvananthapuram

Dr. Mathew Abraham,
Professor neurosurgery,
Department of Neurosurgery,
SCTIMST,
Thiruvananthapuram



Dr. Easwer H.V.,
Professor and head,
Department of Neurosurgery,
SCTIMST,
Thiruvananthapuram

ACKNOWLEDGEMENT

The guidance of Prof. Easwer HV, Professor and Head of the Department of Neurosurgery, has been invaluable and I am extremely grateful and indebted for his contributions and suggestions, which were of invaluable help during the entire work. He will always be a constant source of inspiration to me.

I owe a deep sense of gratitude to Dr. Ganesh Divakar for his invaluable advice, encouragement and guidance, without which this work would not have been possible. His critical remarks, suggestions, helped me in achieving a high standard of work.

I am deeply indebted to Dr. Mathew Abraham, Dr. Krishnakumar K., Dr. George Vilanilam, Dr Jayanand Sudhir, Dr. Prakash Nair, Dr Tobin George, and colleagues and I thank them for their constant encouragement and support.

I owe a deep sense of gratitude to all my patients without whom this work would not have been possible.

I am very grateful to my family, specially my wife Yogineeba Gohil, Dr Amjad and colleague for their constant encouragement and moral support.

Dr. Jaypalsinh Gohil

INDEX

Chiari malformation:

"Chiari I malformation is defined as the descent of cerebellar tonsils below the foramen magnum evident on imaging. if descent is more than 5 mm for adult and more than 3 mm for a pediatric age group is the cut-off limit for definition. (1)"

The Chiari I malformation (CIM) is the most common type, with a prevalence of 0.56% to 1.00% (1). Between 50% and 60% of CIM, patients are diagnosed with a concurrent condition, most commonly syringomyelia. Scoliosis is common not only in CIM but also in syrinx patients. Syringomyelia is regarded as a possible contributor to the pathogenesis of scoliosis in both CIM and idiopathic syringomyelia patients (2,3). Researchers have previously investigated the characteristics of syrinx and scoliosis in the CIM and idiopathic syrinx (IS) population separately (4) and there is no literature on differences between CIM and IS in terms of syrinx characteristics, scoliosis features, and clinical manifestations.

There are many studies on the outcome of Chiari malformation following foramen magnum decompression. However recently few authors have recommended C1-C2 fusion as an effective treatment of Chiari malformation and questioned the effectiveness of foramen magnum decompression. Our institute harbors the largest operated case number of Chiari malformation. We have done a retrospective analysis of clinical and radiological parameters changes following foramen magnum decompression.

AIM AND OBJECTIVES

The study aims to document the long-term effect of foramen magnum decompression surgery on various radiological and clinical parameters and how they change over time after surgery.

REVIEW OF LITERATURE

Historical aspects:

The first description about a myelodysplastic individual indirectly refers to hindbrain herniation was seen in observational Medicae,1641, by famous dutch physician Nicolaes Tulp. Later in 1829, Jean Cruneilher of Paris described a patient born with myelomeningocele. (2)

The first description of hindbrain herniation without myelodysplasia which is currently known as Chiari malformation I was described by Theodor Langhans of Germany. He also described spinal cord syrinx formation due to pathology at the foramen magnum. (3)

Hans Chiari will be remembered for his exclusive work on hindbrain herniation which is now called Chiari Malformation. He belongs to Vienna but most of his work accomplished while he was in Prague. In 1891 he published his work related to Hindbrain malformation. He described a 17-year-old woman who died of Typhoid fever and suffered from hydrocephalus but there were no symptoms related to the cerebellum or medulla. He described his findings as "*peg-like elongation of tonsils and medial divisions of the inferior lobes of the cerebellum into cone-shaped projections, which accompany the medulla oblongata into the spinal canal while sparing the medulla.*"(4)

1894: Julius Arnold described a single case report of the myelodysplastic infant without hydrocephalus associated with the descent of the 4th ventricle and cerebellum, sparing Medulla. (5)

This description was similar to Chiari Type II malformation, however, later Chiari refined his work as a "*displacement of part of the lower vermis, displacement of the pons and displacement of the medulla oblongata into the cervical canal and elongation of the fourth ventricle into the cervical canal"*

In 1907, the Chiari II malformation was renamed Arnold Chiari's malformation by Schwalbe and Gredig to immortalize his professor, Arnold. However in the end it was a significant contribution of Chiari who shed the light on this malformation, Hence now we are referring to it as Chiari II malformation. (6)

In 1883, Cleland noticed the type II malformation but labeled it as Basilar Impression but it contributed little in understanding this malformation. (7) Chiari described Type III malformation which is a lethal one, as the absence of cerebellar tentorium with prolapse of the fourth ventricle and cerebellum into the cervical canal.

Surgical history:

The first attempt at surgical correction was done by Van Houweninge Graftdijk in his thesis entitled over hydrocephalus in 1930. He mentioned "I decided to try widening of the space through which brain had herniated in order to improve CSF flow." however this patient died. This was the first unknown attempt at surgical correction of hindbrain herniation. (8)

In 1938, Penfield has reported a 29-year-old female with hearing loss and right-side face weakness with a past history of thoracic spina bifida surgery in childhood. She underwent posterior fossa decompression with a clinical diagnosis of acoustic neuroma in mind, however, the patient survived for 2 months and later died. On autopsy finding Chiari II malformation with hydrocephalus was revealed. (9)

Later in 1938, McConnel and parker reported good outcomes in 2 out of 5 patients who underwent posterior fossa decompression**.** (7)

In 1948 Chorobski and Stepien reported improvement in symptoms after posterior fossa decompression for Valsalva induced headache. (3)

In 1950, Gardener reported improvement in 13 patients out of 17 patients after posterior fossa decompression and plugging of obex for syringomyelia. After his publication, many adopted posterior fossa decompression as a modality of treatment for hindbrain herniation with syringomyelia. (5) There are more than 1500 publications after 1950 for the management of Chiari malformation. (10)

Chiari, I, and II malformation have been reported with the association of many craniofacial, vertebral, and other CNS anomalies. The few are described below. For an understanding of the pathophysiology of Chiari malformation various theory exists. however, there is no single theory that explains the pathological mechanism for hindbrain herniation various theories pertinent to hindbrain herniation pathophysiology given below.

Table 1: Various associated anomalies with Chiari malformation

Cranium bifidum	Is the cranial counterpart of spina bifida and is likewise comprised of occulta, cystica, and aperta variants	Padget (11)
Lacunar skull	Is characterized by rounded (punched-out) defects in the inner table of the skull separated by a whorl-like bony ridge	Ingraham and Scott (12);
Platybasia	Flattening of the angle between clivus and anterior basicranium; severe form is associated with basilar invagination	Schady et al. (13); Smoker (14)
Small posterior cranial fossa	Reduction in the size of posterior cranial fossa concerning the cranial dimensions	Schady et al. (13)
Basilar invagination	Abnormal approximation of the odontoid process and skull base	Schady et al. (13)
Atlantooccipital assimilation,Atlantooccipital assimilation	Partial or complete fusion of atlas and occipital bone; is seen in about 8 % of pediatric patients with Chiari I malformation, and The fusion of two or more cervical vertebrae; is seen in about 3 % of patients with Chiari I malformation	Tubbs et al. (15)

Spina bifida	Is composed of spina bifida occulta, cystica, and aperta variants; the latter two are comprised of meningocele, meningomyelocele, myelocystocele, and myeloschisis; is often but not always associated with hydrocephalus and Chiari II malformation	Russell and Donald (9); Ingraham and Scott (12)
Hydrocephalus	Often communicating; maybe a primary event or secondary to hindbrain herniation	Ingraham and Scott(12)
Large massa intermedi	Excessive approximation and adhesion of the thalami and thickening of the interthalamic adhesion	Gardner (16)
Stenosis of aqueduct of Sylvius	It May be primary or secondary to midbrain compression by hydrocephalus or overcrowded brain	Masters (17); Russell and Donald (9)
Tectal beaking	The quadrigeminal plate of the midbrain is fused into a conical mass, the apex of which projects between the cerebellar hemisphere	Peach (18)
Dorsal wedging of the brain stem	The dorsal part of the pons and/or upper medulla protrudes into the fourth ventricle	Lichtenstein (19)

"Tight" cisterna magna	The cisterna magna is small or obliterated; is due to overcrowding of the posterior cranial fossa, downward displacement of the cerebellum, fibrovascular adhesions of the meningeal layers, or dysgenesis during embryonic and early fetal periods	Gardner (20);
Low-lying and obliterated fourth ventricle	The fourth ventricle is slit-like and compressed and partly or entirely extends below the foramen magnum	Russell and Donald (9)
Syringomyelia	The cavitation within the spinal cord tissue; is more common in Chiari II than in Chiari I malformation	Josef and Fehlings (21)
Hydromyelia	Dilated central spinal cord canal	Ingraham and Scott (12)

Table 2: Theories for Chiari malformation development

Hydrocephalic Brain or Pressure Coning Theory	Primary agenesis of foramina at the roof of the fourth ventricle causes hydrocephalus and hindbrain herniation resulting in further obstruction and a vicious cycle.	Hans Chiari (8,22)
External Compression Theory	During early fetal life, fistulous communication between the neurocele and amniotic cavity transmits pressure into the cranium and herniation of the hindbrain.	Cameron (23)
Posterior Cranial Fossa Overcrowding in Chiari Malformation	Hydrodynamic theories and obstruction of 4th ventricle outlet	Gardner (24)
Occipital Dysplasia Theory	Underdevelopment of basiocciput	Marin-Padilla (25)
The Neural Tube Overgrowth or Disorganized Neural Tube Growth Theory	Local overgrowth causes spina bifida and overgrowth of the hindbrain causes Chiari malformation	Barry, Patten, and Stewart (26)
Neuroschisis Theory	During development, due to genetic or environmental factors causes fusion of the neural tube wall results in hydrocephalus and small posterior fossa, and hindbrain herniation.	Padget (11)

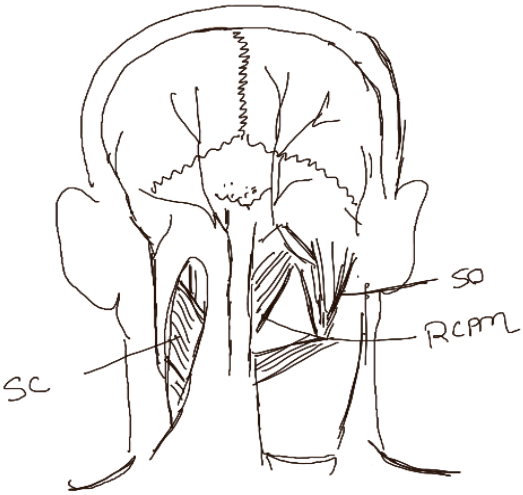
Cord Traction or Tethered Cord Theory	spinal cord fixation at the level of the bifid vertebral arch produces traction on the cord during vertebral growth. This traction interrupts the normal ascent of the spinal cord and results in downward traction of the brain stem and spinal cord and nerves above the point of fixation.	Penfield and Coburn (27)
DevelopmentalArrestTheory	Chiari malformations are a consequence of failure in the normal development of the pontine flexure.	Daniel and Stritch (28)
Inadequate Ventricular Distension Theory	Due to persistent neural tube defects and inadequate pressure, the cranial vesicle fails to expand and results in the smaller posterior fossa. Subsequent Rhombencephalon development causes downward displacement of the brain stem and cerebellum.	McLone and Knepper (29)

Craniocervical Growth Collision or Caudocranial (Reversed) Vertebral Growth Theory	With fixation of the spinal cord to the caudal vertebrae, the upward ascent of the cord is restricted. Therefore, skeletogenic materials are distributed cranially, colliding with the develop- ing skull base. This pattern is referred to as a caudocranial direction of vertebral growth (reversed cervical growth or reversal of craniocaudal vertebral growth), which, according to Roth, leads to various anomalies seen in Chiari malformation.	Roth (30)
Theory of "Suck and Slosh" Effect as the Cause of Origin and Expansion/ Maintenance of a Spinal Cord Syrinx	Intracranial-intraspinal pressure dissociation	Williams (31)
Exaggerated Spinal CSF Systolic Wave Theory for Syringomyelia	Systolic pressure causes fluids in the subarachnoid space to move into the spinal cord secondary to tonsillar ectopia	Oldfield (32)

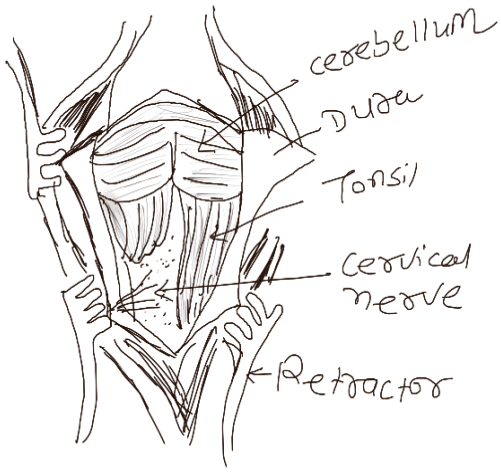
Figure 1

A

B



C



D

Anatomy:

Soft Tissues of the Posterior Craniocervical Junction:

Advancing through the soft tissues overlying the Craniocervical junction, a standard midline incision through the skin advances through the tela subcutaneous and then the upper fibers of the trapezius muscle inserting on either side of the occipital bone near the most posterior aspect of the external occipital protuberance referred to as the inion.

Deep to the trapezius muscle is the splenius capitis muscle, which has a fiber course that is opposite to that of the overlying upper fibers of the trapezius muscle.

The splenius capitis arises from the lower half of the nuchal ligament and spinous processes of the last cervical and upper three to four thoracic vertebrae and attaches to the mastoid process and lateral aspect of the superior nuchal line.

Below the splenius capitis lies the semispinalis capitis muscle (Figure 1 c) which at this point is composed of vertical fibers traveling cephalad to insert onto the occipital bone between the superior and inferior nuchal lines.

Along with their bony attachments, the above-noted muscles also travel along and attach to the midline nuchal ligament, which is a continuation of the supraspinous ligament of the thoracic spine.

As the deepest muscular layer, the suboccipital muscles are four in number. Of these the suboccipital triangle is composed of the rectus capitis major and inferior capitis inferior and superior (Figure 1B). The rectus capitis minor is found just medial to the rectus capitis major attaching the posterior tubercle of the atlas to the occiput. For the suboccipital triangle muscles, the rectus capitis major travels between the occiput and spinous process of C2. The superior oblique arises from the transverse process of the atlas and inserts onto the occiput. The inferior oblique originates from the spinous process of C2 and attaches to the transverse process of the atlas.

Except for the trapezius, which receives its innervation via the spinal accessory nerve, the other above-noted muscles are all innervated by adjacent dorsal rami of spinal nerves.

In unison, the trapezius, splenius capitis, and semispinalis capitis aid in extending the neck. The suboccipital triangle muscles and rectus capitis posterior minor although theoretically, these may move the atlas and axis, practically they may act more as proprioceptive structures. This assumption is strengthened by the fact that many of these small muscles are removed with posterior fossa decompression, and in our experience, no patient has ever complained of a functional deficit postoperatively that would be related.

Vascular structure:

Vascularly, in this region, and from superficial to deep, one finds branches to the overlying skin and muscles derived from the transverse cervical branch of the

thyrocervical trunk which supplies the trapezius muscle, branches of the occipital artery, a posterior branch of the external carotid artery, muscular branches of the vertebral artery, the first branch of the subclavian artery, and segmental branches feeding spinal nerve segments.

The third part of the vertebral artery can be localized as of the contents of the suboccipital triangle.

Posterior Cranial Fossa

The posterior cranial fossa is limited inferiorly by the occipital and sphenoid bones. The posterior aspect of the temporal bones makes up the lateral walls of the posterior cranial fossa (figure 1A) Overlying these bones making up the inferolateral walls is a layer of dura mater, which extends superiorly to form one of the many intracranial Dural specializations, the tentorium cerebelli, which acts as the non-ridged roof of the posterior cranial fossa.

Venous Sinuses:

The mostly valve lacking vertebral venous plexus (Batson's) communicates superiorly with the marginal sinus. This sinus encircles the foramen magnum and receives, among others, the basilar venous plexus anteriorly, veins traveling through the hypoglossal canal laterally, and the occipital sinus posteriorly. This latter sinus will often be encountered in midline decompression of the posterior fossa and is usually enlarged in children. Superiorly, it joins the torcula Herophilus. Interestingly, in the upright position, most intracranial blood travels via the marginal sinus and then into the vertebral venous plexus and through the internal jugular veins in recumbence.

The basilar venous plexus also communicates with the inferior petrosal sinus that then drains into the internal jugular vein either intra- or extracranially. The superior petrosal sinus unites the cavernous sinus anteriorly to the transverse sinus posteriorly and runs in the attached edge of the tentorium cerebelli, which will often house several venous lakes.

The paired transverse sinuses connect venous blood flowing into the torcula Herophilus from, for example, the superior sagittal and straight sinuses to the sigmoid sinuses, which after a short course, drain into the superior jugular bulb.

Dural Innervation

The innervation of the dura mater of the posterior fossa is important as many pain symptoms found in patients with Chiari malformations can be explained due to irritation of such nerves. The dura mater of the cranium is a two-layered membrane that is derived from neural crest cells. However, the spinal dura mater is a single layer and is derived from the paraxial mesoderm. This difference in derivation helps one understand the varied innervation pattern seen between the posterior fossa and cervical dura mater. In the cervical spine as well as the remaining spinal dura, the recurrent nerve of Luschka (recurrent meningeal or sinuvertebral nerve) segmentally innervates the dura mater. These meningeal branches arise from the spinal nerves and enter the intervertebral foramen to innervate the dura as well as the adjacent annulus fibrosis, periosteum, and posterior longitudinal ligament.

The dura mater of the posterior cranial fossa receives multiple nerves that contribute to its innervation. Branches have been found to arise from the facial, glossopharyngeal, vagus, and hypoglossal nerves. The majority of fibers from the hypoglossal nerve are thought to arise from the upper cervical nerves.

Once overlying muscles have been dissected away, the posterior atlanto-occipital membrane can be observed. This structure is often thickened in the Chiari malformation and travels between the posterior arch of C1 to the posterior aspect of the occiput. A venous plexus may be found within this membrane and the third part of the vertebral artery will pierce it before traversing.

Posterior Aspect of the Atlas

The posterior arch of the atlas is easily seen with its midline posterior tubercle for the attachment of the rectus capitis posterior minor. The posterior arch may be bifid or assimilated to the occiput (atlantooccipital fusion). Lateral on the posterior arch,
the horizontal segment of the vertebral artery, following leaving the transverse foramen of the atlas, courses around the superior articular process to pierce the posterior atlanto-occipital membrane. A bony foramen (arcuate foramen) may be found at this location. The periosteum along the anterior surface of the posterior arch of the atlas may be thickened in the Chiari malformations.

Intradural Anatomy of the Craniocervical Junction (fig 1A, 1B, and 1D)

First Denticulate Ligament

Intradural at the Craniocervical junction and from a posterior perspective, some of the lower cranial nerves and upper cervical nerves are observed. A good landmark for these structures is the first denticulate ligament, which is a pial extension from the C1 segment of the spinal cord to the inner aspect of the intracranial dura mater. Superiorly, this ligament attaches near the entrance of the third part of the vertebral artery, and inferiorly, it separates the vertebral artery and ventral rootlets of the upper cervical spinal nerves anteriorly from the dorsal rootlets of the upper cervical spinal nerves and spinal accessory nerve posteriorly. Of note, the dorsal roots are not always present nor are the dorsal root ganglia of C1. The ventral root of the C1 spinal nerve is often connected to the spinal accessory nerve via a communicating branch.

Accessory Nerve

Although variation exists, the spinal accessory nerve arises from the upper five or so cervical spinal cord segments, and connections between the nerve and especially the dorsal rootlets of the upper cervical spinal cord segments are common. The spinal accessory nerve will ascend from its spinal origin to enter the foramen magnum and, near the jugular foramen, unite with its cranial part to exit the skull.

Hypoglossal Nerve

Just superior to the uppermost attachment of the intracranial and first denticulate ligament is the hypoglossal nerve, which will innervate three of the four extrinsic tongue muscles.

First Spinal Nerve

The C1nerve, as mentioned above, travels posteriorly to innervate the muscles of the suboccipital triangle including the rectus capitis posterior minor and overlying semispinalis capitis muscle. This nerve, in general, does not have a cutaneous distribution. Anteriorly, the ventral ramus of the C1 spinal nerve contributes to the cervical plexus and travels along the hypoglossal nerve to terminate on the thyrohyoid and geniohyoid muscles.

Posterior Spinal Artery

The posterior spinal arteries arise from the vertebral arteries and travel around the brainstem and then inferiorly along the posterolateral surface of the cervical spinal cord. They are distinct single vessels only at their origin and distally become irregular anastomosing channels, retaining, to a large degree, their embryonic plexiform pattern. The posterior spinal arteries are the largest in the cervical and lumbar regions.

Posterior Inferior Cerebellar Artery:

An important artery of the posterior fossa, especially regarding the Chiari malformation, is the posterior inferior cerebellar artery (PICA), which arises from the vertebral artery near the vertebrobasilar junction and travels inferiorly toward the foramen magnum.

PICA can be divided into five segments:

1. An anterior medullary segment, which is often absent (i.e., PICA does not originate anterior to the medulla oblongata), extends from the origin of PICA to the inferior olive.

2. A lateral medullary segment that extends from the inferior olive to the origins of the lower cranial nerves.
3. A posterior medullary segment (tonsillo-medullary segment) begins where the PICA passes posterior to the lower cranial nerves and ends where the ascending vessel reaches the midlevel of the medial surface of the tonsil. It passes immediately posterior to the roof of the lower half of the fourth ventricle. All medullary segments give rise to perforating branches, which, if injured, are the reason nuclear dysfunction occurs (e.g., Wallenberg syndrome).
4. A supratonsillar segment (telovelotonsillar segment) begins at the midportion of the tonsil, includes the cranial loop, and ends where the PICA exits the fissures between the vermis, tonsil, and cerebellar hemisphere to reach the suboccipital surface. This is the most complex of the PICA segments.
5. Cortical segments (hemispheric segments) supply such areas as the midline vermis and tonsils. The PICA often bifurcates into medial and lateral trunks where the vessel emerges onto the inferior cortical surface. The medial trunk gives rise to vermian and tonsillar branches, and the lateral trunk gives rise to hemispheric branches.

Cerebellar Tonsils

The cerebellar tonsils (figure 1 a and 1b) are normally found several millimeters above the foramen magnum and are often asymmetric between the left and right sides. These structures are connected to the cerebellum along their upper lateral surface by the so-called tonsillar peduncle. Laterally, the cerebellar tonsils are covered by the biventral lobule. The cerebellomedullary fissure separates the tonsil from the posterior surface of the medulla oblongata. The space that separates the left and right tonsils across the

midline is referred to as the vallecula. At its superior pole, each tonsil's anterior surface faces the nodule, inferior medullary velum, and tela choroidea. This superior pole of the tonsil, which faces the uvula medially, is separated from the aforementioned structures by an extension of the cerebellomedullary fissure known as the telovelotonsillar cleft.

Retrotonsillar Veins

The superior retrotonsillar vein originates from the superior pole of the cerebellar tonsil and travels posteriorly to unite with the inferior retrotonsillar vein, which arises near the inferior pole of the tonsil to course superiorly. Together, these vessels form the inferior vermian vein, which therefore drains the medial and lateral surfaces of the tonsils. The inferior vermian vein may drain into the tentorial or transverse sinuses or the torcular Herophilus.

Relation to the fourth ventricle:

The foramen of Magendie is a median aperture resting at the inferior aspect of the fourth ventricle and, with the laterally positioned foramina of Luschka, allows CSF egress from the fourth ventricle into the cisterna magna and cervical subarachnoid space. The foramen of Magendie may be stenotic or imperforate as it is in other mammals. Interestingly, neither the foramen of Magendie nor the foramina of Luschka are lined with ependyma.

Classification of Chiari Malformations

I: Tonsillar ectopia ≥ 5 mm into the foramen magnum. No neural tube defects.

II: Neural tube defect. Hy- hydrocephalus and other anomalies.

III: Rare, a portion of the cerebellum and brainstem migrates out of the Craniocervical junction.

IV: Uncommon, hypoplastic or absent cerebellum.

0: Crowded appearance of Craniocervical junctions. No or minimal tonsillar ectopia. altered CSF flow on Cine MRI.

1.5: Significant caudal descent of the cerebellar tonsils and brainstem. No neural tube defects.

Genetics:

The accumulation of data over the years has established that there is a preponderance of evidence supporting a genetic contribution to at least a subset of Chiari I malformation patients, and the field is at a point where significant progress is being made in this area of research. Although there is strong evidence that genetics plays an important role in the development of at least certain forms of Chiari malformation, Chiari malformation is likely to have a multifactorial etiology, influenced by genetic, epigenetic, as well as environmental factors. While it is important not to discount these other nongenetic contributions to disease, there are many potential benefits of genetics research. Future genetic analyses will likely result in the identification of a gene or genes that increase susceptibility to Chiari malformation that may later be translated into a genetic test resulting in more accurate and quicker diagnoses. This can be especially useful for complex diseases, such as Chiari malformation, where the symptoms are vague and not unique to a disorder, resulting in slow diagnoses and even misdiagnoses. (33) Understanding the genetics of disease can provide more information on the underlying disease mechanism and can also indicate which specific biological processes play a role in disease development. There is also the exciting potential for the development of new therapies and treatments targeting identified genes or pathways

that are dysregulated, which may ultimately provide patients with alternative treatment options to surgery. (33–35)

Syringomyelia pathogenesis:

Syringomyelia is characterized by tubular cavitation of the spinal cord that may extend over many segments. The cavity is typically filled with clear fluid that closely resembles cerebrospinal fluid (CSF) or extracellular fluid (ECF) (36,37).

Hydromyelia is defined as dilatation by the fluid of the central canal of the spinal cord. It is frequently seen in association with some forms of the syrinx and can be indistinguishable, even at autopsy, from syringomyelia (38)

Syringobulbia is rare but strongly associated with syringomyelia and connotes fluid-filled or slit-like cavities in the brainstem, most commonly the medulla (39)

Classification systems vary but most divide syringomyelia into two broad categories, communicating and noncommunicating. Communicating (hydromyelic) syringomyelia is often associated with Chiari type II malformations and is an ependyma-lined expansion of the central canal that is contiguous with the fourth ventricle (40–42) complex hindbrain malformations such as encephalocele, Dandy-Walker variants, hydrocephalus secondary to haemorrhage or meningitis, or other conditions that increase intracranial pressure can fall within this category. Acquired forms are typically

bound caudally by central canal stenosis, a normal age-related phenomenon in humans (40)

Noncommunicating (syringomyelic) syringomyelia is more common and characterized by complex cavitation that does not communicate rostrally with the fourth ventricle. These syringes may involve both the ependyma-lined central canal and paracentral spinal cord parenchyma where the syrinx demonstrates a gliotic lining. Chiari malformations (type I and type 0) are commonly characterized by syringes of this type (42)

Other etiologies in this category could include syrinx secondary to spinal arachnoiditis (postmeningitic or posttraumatic), subarachnoid hemorrhage, extramedullary compressions (cyst, tumor, or spondylosis), and other skeletal abnormalities such as basilar impression. Noncommunicating syringomyelia can also include primary parenchymal cavitation such as trauma, intramedullary haemorrhage, or infarction (42)

Proposed Theories of Syringomyelia Formation

Gardner's Hydrodynamic/ Water-Hammer Theory

In 1959, Gardner and Angel suggested that syringomyelia forms because of a persistent opening of the central canal at the obex, in the setting of closed fourth ventricular outlet foramina (37). This hypothesis was the basis upon which plugging of the obex was suggested as part of the treatment for syringomyelia. Subsequently, Gardner expanded on his theory to propose that syringomyelia is a result of direct transmission of a CSF pulse through the obex in a "water-hammer" fashion (37)

Inconsistencies of Gardner's Theory

Gardner's theory could not explain the following observations: First, if one were to assume that the pathophysiology of syringomyelia is invariable regardless of etiology, the hydrodynamic theory can- not explain cyst formation secondary to trauma, arachnoiditis, tethered cord, etc. Second, this single theory of pathogenesis at the foramen magnum does not account for the syrinx septations that are often evident on MRI. Third, West and Williams showed using ventricular contrast studies that the obex is patent in only 10 % of patients, thus refuting Gardner's hypothesis. (31,36,43)

Williams' Modifications of Gardner's Hydrodynamic Theory (The Suck Effect Theory):

Based on manometric observations in normal subjects and Chiari I patients, Bernard Williams devised a theory that examined syringomyelia from another perspective (31). Like Gardner, he postulated an obstruction at the foramen magnum. However, he theorized that the Chiari I malformation is an acquired anomaly that results from the excessive molding of the head, perhaps during delivery through the birth canal, which may then cause hindbrain adhesions and related outlet obstruction.(44,45)

In support of this claim, he showed using ventricular contrast that posterior fossa arachnoiditis correlates strongly with a history of a difficult birth. Williams hypothesized that hindbrain adhesions can result in transient pressure differentials between the cranial and spinal compartments due to epidural venous congestion, particularly during Valsalva manoeuvres (coughing, sneezing, straining). This, in turn,

may cause a delay of caudad CSF flow while maintaining normal craniad flow, and as a result, fluid is "sucked from the ventricle into the central canal."(31,44)

Perivascular CSF Dissection Theory:

Oldfield et al. expounded on the perivascular CSF dissection theory by providing favourable observations using various imaging studies. They showed that the rostro caudal movement of the spinal cord results in the CSF dissection in the subarachnoid space and documented such movement both intraoperatively using ultrasonography, as well as on dynamic MRI studies. (46)

Unlike Williams' theory, in which the CSF dissection is driven by Valsalva manoeuvres, Oldfield et al. proposed that normal CSF pulsations provide a more or less continuous reason for fluid to enter the spinal cord. They specified that the displaced cerebellar tonsils act as a piston as they are propelled caudally with systole, thus creating a pressure wave within the entrapped subarachnoid space and syrinx. One might consider as supportive evidence of the perivascular CSF dissection mechanism, and specifically Oldfield's piston effect theory, the recent observation of a "syrinx state," in which spinal cord oedema precedes syringomyelia (39,47)

Intramedullary Pulse Pressure Theory:

Based on animal experiments, Greitz' group developed a theory that suggests that the fluid within a syrinx derives from extracellular fluid forced into the spinal cord from a high-pressure microcirculation rather than high-pressure CSF from the spinal cord subarachnoid space (48)

Specifically, they state that when the subarachnoid space is obstructed from any cause (Chiari I, tumour, arachnoiditis, etc.), there is a significant decrease in pressure transmission to the distal CSF spaces and concomitant increased transmission of the systolic CSF pulse pressure into spinal cord parenchyma close to the obstruction. This imbalance of pressures between the spinal cord and subarachnoid space leads to distention of the spinal cord just below the blockage. Furthermore, part of the systolic CSF pulse pressure is "reflected" into the spinal cord at the site of obstruction, also distending the spinal cord above the blockage. (37,49)

despite tremendous advancements in technology and considerable effort by many researchers, no single theory has so far definitively solved the enigma of Chiari-related syringomyelia formation. However, the intramedullary pulse pressure theory seems to best explain the formation of syringomyelia independent of etiology.

Radiology:

The imaging study of choice to evaluate patients with cerebellar ectopia is MRI. Sagittal and axial T1- and T2-weighted MRI with images no thicker than 3 mm should be obtained in all patients. The images should include the entire brain and skull, as it is important to look for hydrocephalus, masses, malformations, deformities, or evidence of high or low intracranial pressure. The cervical spine should also be evaluated down to C6–C7 to look for associated spinal cord edema or syringohydromyelia; as it may be difficult to differentiate a syrinx from edema on T2 sagittal images, axial T1 images are useful for confirmation. The key anatomic imaging finding on MRI of these malformations is tonsillar ectopia with compressed ("peg-like" or "pointed") cerebellar

tonsils and nearly complete effacement of CSF at the foramen magnum or C1 level. For adult displacement, more than 5 mm below the foramen magnum and pediatric more than 3 mm displacement are considered significant. (33)

The imaging study might also show hydrocephalus or a posterior fossa mass causing cerebellar herniation through the foramen magnum. In patients with diseases of bone, basilar invagination may result in tonsillar herniation.

CSF flow study:

Normal studies demonstrate craniocaudal CSF flow (white CSF) during cardiac systole, which lasts about 40 % of the cardiac cycle, (50) the lower brain stem and cerebellar tonsils move slightly downward. During cardiac diastole (60 % of cycle), CSF moves rostrally and is black; the brain stem and tonsils move slightly upward. (50) When the CSF flow is impaired by cerebellar tissue in the foramen magnum, the amount of CSF seen in motion ventral and dorsal to the brain stem and the craniocervical junction is reduced, CSF systole is shortened, diastole is prolonged, and movement of the brain stem and tonsils is increased (51,52). When flow studies demonstrate prolonged CSF diastole at the foramen magnum with an increased downward motion of the cerebellar tonsils and brain stem, it is strong evidence of impaired CSF flow.

Chiari II

For assessment of the Chiari II malformation, the complex set of the posterior fossa, and supratentorial abnormalities seen in the setting of myelomeningocele, MRI again forms the diagnostic mainstay. However, prenatal screening ultrasound is often where the brain and spine abnormalities in the Chiari II malformation complex are identified

initially and deserves discussion in this setting. Subsequent fetal MRI is indicated for further characterization and identification of associated findings. The appearance of myelomeningocele and the Chiari II malformation on prenatal imaging will be commensurate to the relatively early developmental stage, with abnormalities often progressing on follow-up studies. Maternal serum screening tests or amniocentesis, performed within the same period, revealing elevated alpha-fetoprotein in the setting of open neural tube defect can also be the referring indication for fetal brain and spine MRI. After delivery, MRI is the study of choice for children with myelomeningocele.

The posterior fossa is small with little or no CSF space. The tentorium cerebelli has a low attachment and steep orientation, causing the straight sinus to have a nearly vertical course to the torcular Herophili. The cerebellum nearly fills the small posterior fossa with hemispheres wrapping laterally around the brain stem. The fourth ventricle is small and narrowed in its rostrocaudal dimension. The pons may be pushed against the clivus, and the pressure of the hindbrain structures on the skull base can cause concave erosive changes upon that structure. The foramen magnum and tentorial incisura are enlarged due to the chronic herniation of the cerebellum through these structures. The cerebellar hemispheres and vermis are often very small possibly due to chronic ischemia from compression.

The inferior colliculi are often enlarged and stretched in an inferior and dorsal direction, while the superior colliculi are small, resulting in the “beaked tectum” appearance. The supratentorial vault is small, again, most likely due to inadequate ventricular distension earlier in gestation; however, disproportionate ventricular dilation is common from

narrowing of the aqueduct, posterior fossa crowding, or perhaps lack of dampening of CSF pulsations in the spine because the foramen magnum is occluded (53).

Supratentorial anomaly:

The anterior commissure is in a low position (halfway between the optic chiasm and the foramen of Monro) in 40 %. The corpus callosum has a very variable appearance. Most typically, it is thin in the posterior aspects (posterior body and splenium), similar to what is seen associated with other causes of congenital hydrocephalus. However, ~30 % have frank hypogenesis with absent splenium, absent rostrum, and a small posterior callosal body (54). The Corpus callosum may show hypogenesis or dysgenesis. Anomalies of sulcation are commonly identified. A condition called "stenogyria (narrow gyri)."

Evaluation of the spinal cord differs on the fetal exam as compared to the postnatal (postsurgical) MRI. The purpose of the fetal spine MRI is to identify and characterize the myelomeningocele (MMC) and its levels and to look for associated spine anomalies. (54)

Chiari III

The much more rare Chiari III malformation is also best evaluated with MRI which can characterize the contents of the occipitocervical encephalocele and associated brain and spine abnormalities. Low-dose CT may also help to visualize the bony cervical and/or occipital cranial defects. The purpose of imaging is to define the contents of the encephalocele, including the volume of herniated cerebellum and/ or brain stem,

meninges, and CSF. MR venography is also indicated to assess the location of the dural venous sinuses concerning the mass. These findings are essential for surgical planning and may be helpful for prognostication. (55)

The natural history of Chiari:

The degree of cerebellar tonsillar descent is not always stable over time. The gradual ascent of cerebellar tonsils has been associated with normal childhood development. (56)

There are several reported cases of CM-I that experienced a spontaneous improvement in cerebellar tonsillar descent(57–59)

Any conclusions derived from these asymptomatic or minimally symptomatic patients should not be applied to symptomatic patients that are ordinarily considered good surgical candidates. It seems likely that natural history is worse for more symptomatic patients for whom surgery is more frequently offered

Assessment :

Subjective clinical assessment

The typical CMI symptom is an occipital headache that is short and strain-related (e.g., occurs when coughing, laughing and exercising) (33). This might be related to impaired cerebrospinal fluid (CSF) dynamics and crowding at the foramen magnum or pressure at the meninges. (60) Atypical types of headaches, such as those with a long-duration or that are migraine-like, have been reported, and these symptoms can make further decisions regarding patient management difficult (61). Other common presentations of CMI include ophthalmological manifestations, such as double or blurred vision (33,62).

Furthermore, CMI patients, especially in cases of the associated syrinx, can present with medullary symptoms, such as a loss of sensation or limb weakness (15,33,63). Finally, reports have described CMI patients who presented with unsteadiness and positional dizziness (33). Other atypical presentations of CMI, such as swallowing difficulty or cognitive dysfunctions, have been described in case reports and series. summarizes the proposed origins of these symptoms and the relevant involved structures.

CMI symptoms usually have a gradual onset. However, an acute presentation has been reported in 1.2 to 3.2% of cases (64,65). The clinical presentation of reported acute cases has involved medullary signs and even sudden death (65,66). The risk factors that might help to predict these incidents remain unknown, and evidence is currently based solely on case reports and series.

Neurological examination

In CMI patients, a detailed bedside examination will complement the patient's history and helps when forming a comprehensive assessment before making a final diagnosis. It is also valuable for choosing further - investigational tools if necessary. The cranial nerve problems most frequently encountered in CMI are diplopia and nystagmus (especially downbeat nystagmus), which are characteristic of craniocervical junction abnormalities. (67,68) Cerebellar signs, such as truncal ataxia, have been reported in 40% of CMI cases. (69) These occur in addition to long tract manifestations, such as paresthesia or motor disturbances in the upper or lower limbs. (33,63) Because it is a central lesion, syrinx frequently affects the spinothalamic tracts and causes dissociative

paresthesia (70), which results in the loss of pain and temperature sensation without the loss of fine touch and proprioception. (71)

DIFFERENTIAL DIAGNOSIS

Pseudotumor cerebri (PTC): it can be difficult to differentiate a diagnosis between CMI and PTC. They have a very similar clinical presentation and are more common in females 12,100. However, PTC is highly associated with obesity (72). In the literature, researchers have found that a radiological finding of cerebellar tonsillar ectopia similar to that of CMI is observed in 2-5% of PTC cases (73,74). One study, a series by Banik et al., reported that this number was even higher (24%) (73). Hence, PTC patients may consequently be misdiagnosed as CMI and offered unnecessary OCD, and this could explain why some patients never improve after OCD.

Space occupying lesions

Posterior fossa lesions can displace the cerebellar tonsils caudally towards the foramen magnum due to high ICP, resulting in secondary CMI (75).

Intracranial hypotension

This condition results from low CSF pressure and can develop iatrogenically after lumboperitoneal shunts or lumbar punctures. It is associated with peculiar features on MRI, and these, in addition to the presence of cerebellar tonsillar herniation, aid in its diagnosis. These peculiar features include a sagging brain, meningeal enhancement, and enlarged venous sinuses (76,77)

Basilar invagination

This is one of the most common anomalies of the Craniocervical junction. It is associated with CMI, which is found in approximately 12-13% of cases with basilar invagination. (78) This association has been reported to be even higher, as in Goel, who reported a series of 190 basilar invagination patients, 53% of whom had associated CMI. (79) Its characteristic feature is an inward and upward displacement of the caudal part of the occipital bone to a position close to the vertebral column. Failure to exclude basilar invagination before OCD in CMI might lead to unsatisfactory results and risk of decompensation in affected patients

MANAGEMENT OF CMI

Surgical management of CMI is the treatment of choice in symptomatic cases. Since its first description by Hans Chiari, many operative techniques have been advocated and compared concerning their postoperative outcomes and complications. The first study to describe the use of OCD for CMI was published in 1938 by McConnell and Parker (7).

Choosing which technique will be best in each case is not an easy task, and one should first address whether the CMI is primary or secondary, as previously described. For example, a CMI that is a secondary etiology of cerebellar tonsillar descent may not require OCD. It is, therefore, necessary to rule out any underlying intracranial hyper/hypotension, tumors, craniosynostosis, etc. Part of a basic evaluation involves examining the craniocervical area for any instability (e.g., basilar invagination, which could render OCD a dangerous choice without stabilization and fixation).

Today, the most widely discussed procedures are posterior fossa decompression with duroplasty (PFDD) and posterior fossa decompression without duroplasty (PFD). (80) Besides, other techniques have been performed, including decompression plus resection of the tonsils (81,82), CSF diversion procedures (83,84), the section of filum terminal (85,86), and stabilization/fusion surgeries. (87,88) Strahle et al. reported that both CMI and craniosynostosis were observed in 8% of 383 consecutive patients with craniosynostosis. Almost 50% of these patients had isolated lambdoid synostosis. Additionally, of 7 patients who underwent the only craniosynostosis repair, 6 showed a decrease in tonsillar ectopia, 5 had improved CSF flow, and in 2, a syrinx was resolved. (89) In most reported studies, PFDD was superior to PFD in terms of clinical improvement, syrinx resolution, and reducing the risk of re-operation. However, the risk of postoperative complications was lower and operation times and hospital stays (days) were shorter in PFD than in PFDD. This is not surprising because, in PFD, it is not necessary to open the dura and expose the subarachnoid spaces containing CSF. (80,90–92) Some authors have investigated the impact of MRI flow studies and intraoperative ultrasound **(US)** in this condition. (93) For example, Yeh et al. performed intraoperative US in 130 pediatric patients with CMI, of whom 40 achieved satisfactory US results and therefore underwent only bony decompression. No complications were noted in these 40 patients, whereas complications occurred in 12 of 85 patients who underwent duroplasty, and these patients experienced more postoperative symptoms. Hence, the authors proposed that the intraoperative US could be a valuable tool for choosing an appropriate surgical technique. (94)

The section of the filum terminal is a controversial treatment option. Royo- Salvador proposed that underlying occult tethered cord syndrome is associated with CMI. They

reported performing a section of filum terminale in a small series of CMI patients, and they reported that this group achieved clinical improvement. (85) However, larger and controlled studies with longer-term follow-up are rare and needed to explore this issue. Posterior occipitocervical fixation in CMI can be justified, when feasible, as a necessary approach in select patients, such as those with confirmed instabilities at the craniocervical junction. (95,96) Goel took this further in his published series of 65 CMI patients. He speculated that the primary incident underlying CMI is instability at the C1-2 area regardless of whether the patient had an associated basilar invagination and that, therefore, all CMI patients should be primarily managed with a posterior fixation of C1-2 using lateral mass screws. (21,97) This approach was later criticized because, in Goel's series, the morbidity rate in patients who underwent posterior fixation was higher than the mortality rate in reports of only OCD (1.5% vs. 0.9%, respectively). Also, the risk of injury to the vertebral artery was higher than that observed for conventional CMI procedures. Stabilization procedures are also known to restrict the range of movement at the craniocervical junction. However, it has been suggested that in CMI cases in which stabilization is needed, it is more biomechanically reasonable to perform occipitocervical fixation than atlantoaxial stabilization alone. (98,99)

Surgical complication:

*Intraop*erative *complication*

bleeding,

Compression on brainstem due to hematoma

Lower cranial nerve injury

Incidental plunging

Postoperative complication:

Pseudomeningocele

CSF leak

meningitis

Wound infection

AIMS: The study aims to document the long-term effect of foramen magnum decompression surgery on various radiological and clinical parameters and how they change over time after surgery.

Foramen magnum decompression:

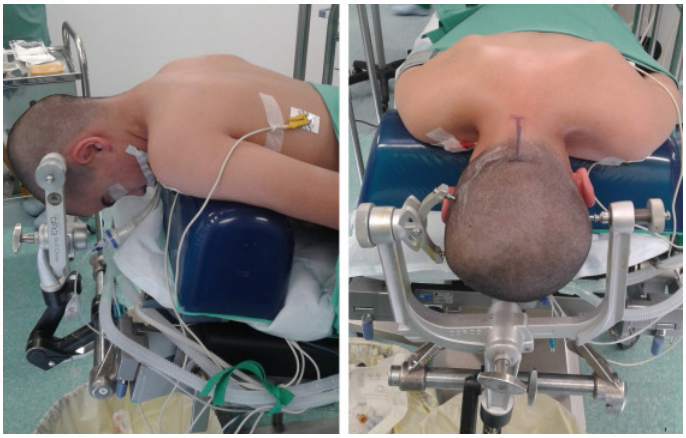


Figure 2A: An illustration of the senior author's position of patient and incision for a midline sub-occipital craniotomy.

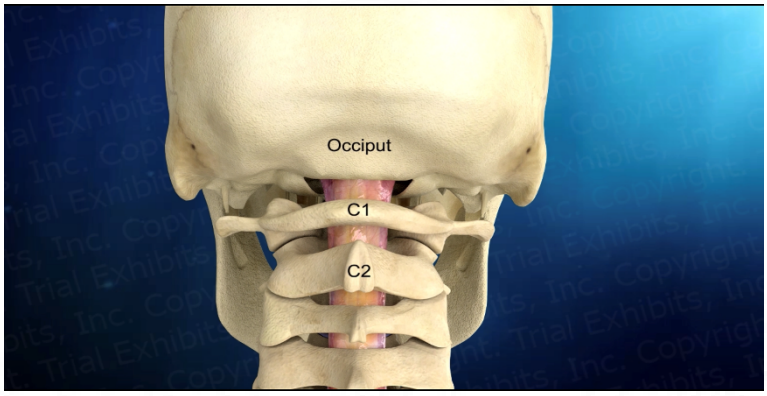


Figure 2B: An illustration of exposure of bone.

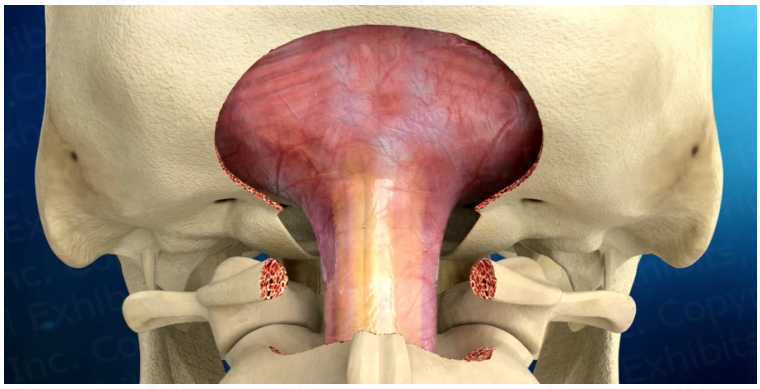


Figure 2C: an illustration demonstrating the dural exposure after suboccipital craniotomy and c1 arch excision.

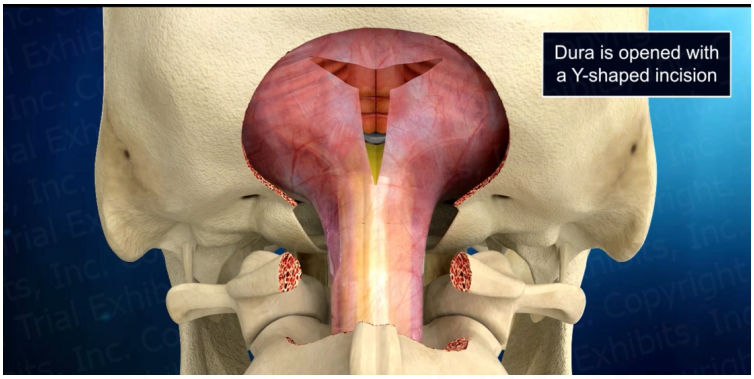


Figure 2D: an illustration demonstrating the dural opening in a Y shape manner.

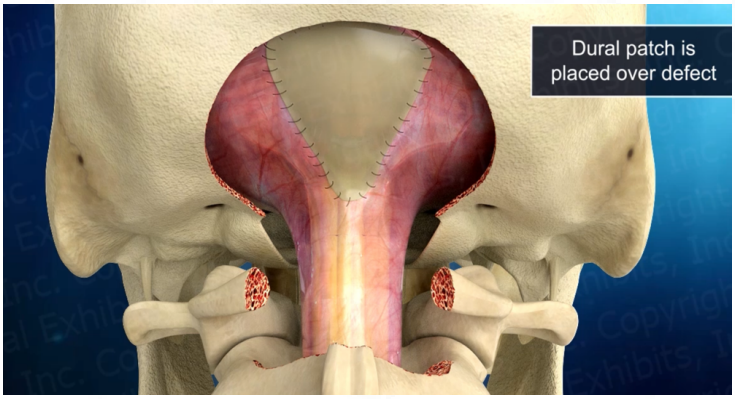


Figure 2E: an illustration demonstrating dural defect repair using a pericranial patch.

MATERIALS AND METHODOLOGY

Study design: a retrospective study is conducted in our institute SCTIMST, Trivandrum, from Jan 20004 to Dec 2019. All symptomatic cases of Chiari malformation with a syrinx and spinal deformity like scoliosis were selected from retrospectively using keywords such as “Chiari malformation”,” Chiari”,” syrinx”,” syringomyelia”,” scoliosis”, “ Chiari malformation with syrinx/syringomyelia” etc.

A total of 728 cases were operated in our institute from the above-defined period. Among this total 101 patients were selected who fulfill selection criteria and whose complete preoperative as well as postoperative clinical and radiological data were available for analysis. Due to the up-gradation of PACS software, many data were not available and such cases were excluded.

Patients: in retrospective analysis, no patient was called or contacted due to the ongoing pandemic.

Research Tool and variables: the primary outcome was measured as changes in syrinx following foramen magnum decompression in Chiari I malformation. The second variable was measured as changes seen in scoliosis parameters following foramen magnum decompression. Tonsillar decent was classified into 3 grade, Grade I -the descent of tonsilar tip below foramen magnum but above C1 arch upper border. Grade II was defined as a descent of tonsilar tip below the C1 arch but above the C2 arch upper border. Grade III was defined as tonsilar descent with the tip at the level or below the C2 arch. T1WI and T2WI sagittal section was used for measurement of the distance of the tonsilar tip from the foramen magnum border. The asymmetry of tonsilar descent was also measured as the difference between the distance of the left

and right tonsilar tip from the foramen magnum. (X-Y, where X left tonsilar tip distance from the foramen magnum and Y is right tonsilar tip distance from foramen magnum). Changes in tonsilar descent in the postoperative period were recorded.

following Syrinx parameters were measured preoperative and postoperative period, observed at the same level using sagittal and axial T2WI MRI. Syrinx diameter and cord diameter at the same level were recorded in both the preoperative and postoperative periods. If syrinx is patchy difficult to measure diameter then postoperative changes were recorded as the same or collapsed or absent.

Clinical parameters: preoperatively age, sex, mode of presentation, duration, comorbidity, motor and sensory deficit, any cranial nerve deficit, cerebellar sign and, presence of nystagmus were recorded. Intraoperative findings such as foramen magnum decompression with or without duroplasty, C1 arch excision, Tonsilar ablation and, arachnoid dissection, etc were recorded. Any neurovascular complications during surgery were recorded. postoperatively any complications such as prolong ventilation, tracheostomy, lower cranial nerve palsy, aspiration, or other cardio-pulmonary complications were recorded. Postoperative improvement in symptoms or deficit on follow up were recorded from postoperative hospital stay charts and follow up OPD visits. If there was any mortality, the cause of death was recorded. If any patients require re-do surgery on follow up, the reason for re-do surgery, and operative findings at the same time were also recorded.

Eligibility criteria:

Inclusion criteria

All operated patients with foramen magnum decompression for CIM without fusion.

Retrospective Consecutive 101 cases will be included

Exclusion criteria:

All recurrent cases will be excluded.

Who underwent Upper cervical fusion

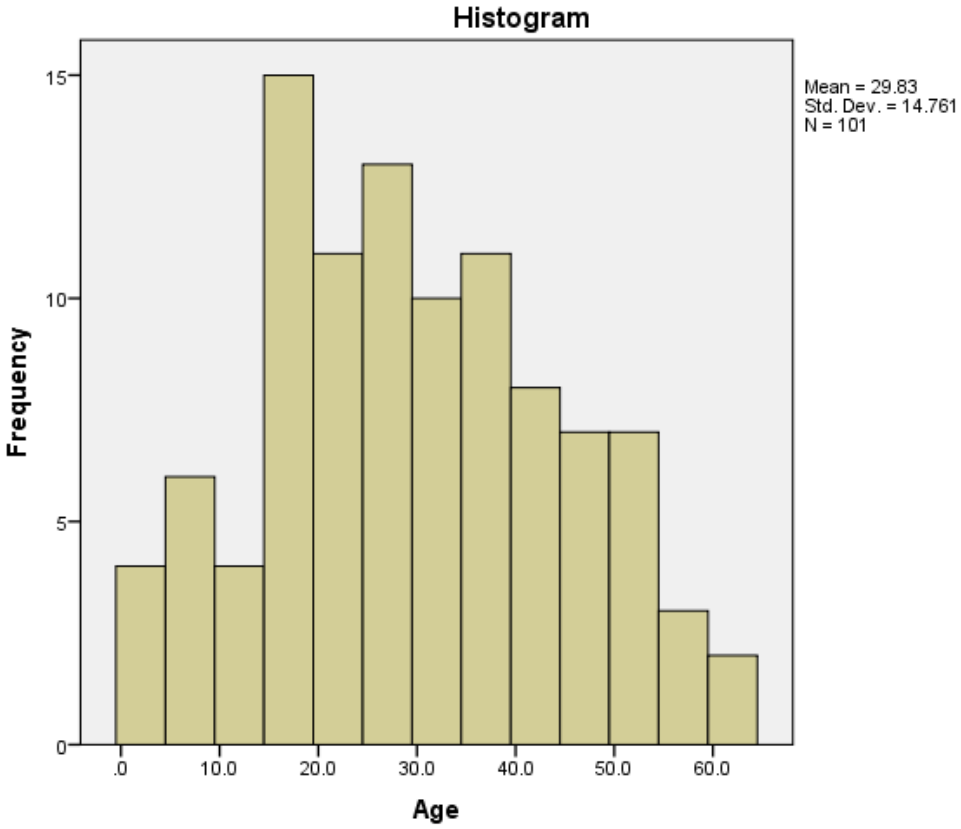
Spinal tumor and Cases with Tethered cord, myelomeningocele

RESULTS ANALYSIS

➢ **Demographic data:**

Since Dec 1998, approximately 728 patients were operated on for Chiari malformation. Radiological and detailed clinical data are available from 2004 onwards. Due to repeated software updates, old patients' radiological data were missing from the electronic system. Out of 728 total, 328 patients were available with radiological and clinical parameters. Out of 328 patients, a total of 101 patients were selected for analysis who fulfilled all selection criteria.

In our study 41 patients (40.6%) were male and 60 patients (59.4%) were female. The minimum age in our study was 2years and the maximum age was 63 years. The mean age was 29.83 years. A histogram of the patient's distribution of various ages is shown below.



➢ **Clinical symptoms:**

Clinical symptoms	Frequency	Percentage
Cough headache	51	50.5
Gait unsteadiness	12	11.9
Sensory disturbance	31	30.7
Posterior column symptoms	21	20.8
Motor weakness	10	9.9

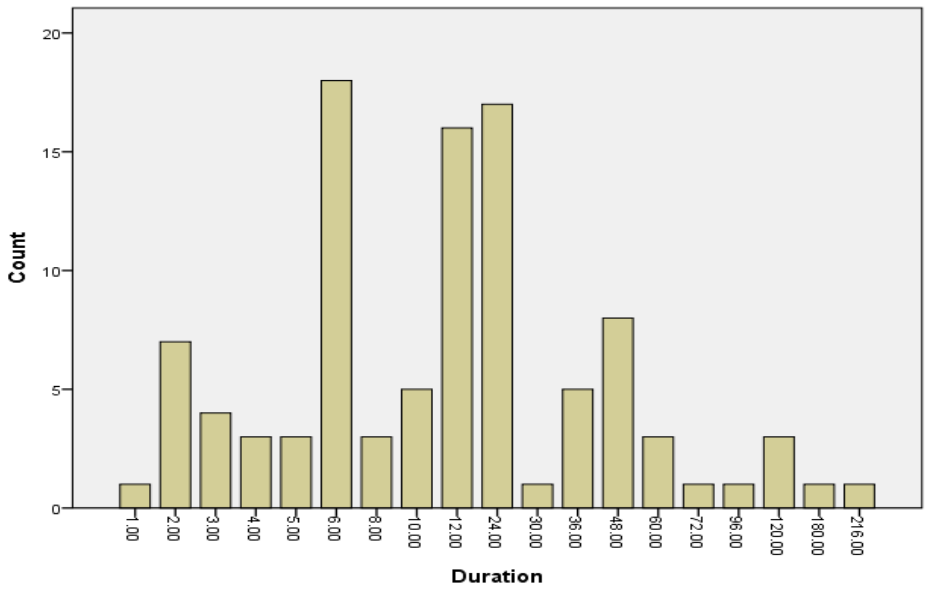
Others	33	32.7

The most common symptom was cough headache. Other symptoms categories include neck pain, autonomic disturbance, clawing of hand, scoliosis, loss of consciousness, back-pain, and urinary symptoms.

- **Duration of symptoms:**

Duration of presentation	frequency	percentage
<=6 months	36	35.6
Between 6-12 months	24	23.8
Between 13-60 months	34	33.7
Between 61-120 months	5	5.0
More than 120 months	2	2.0
Total	101	100.0

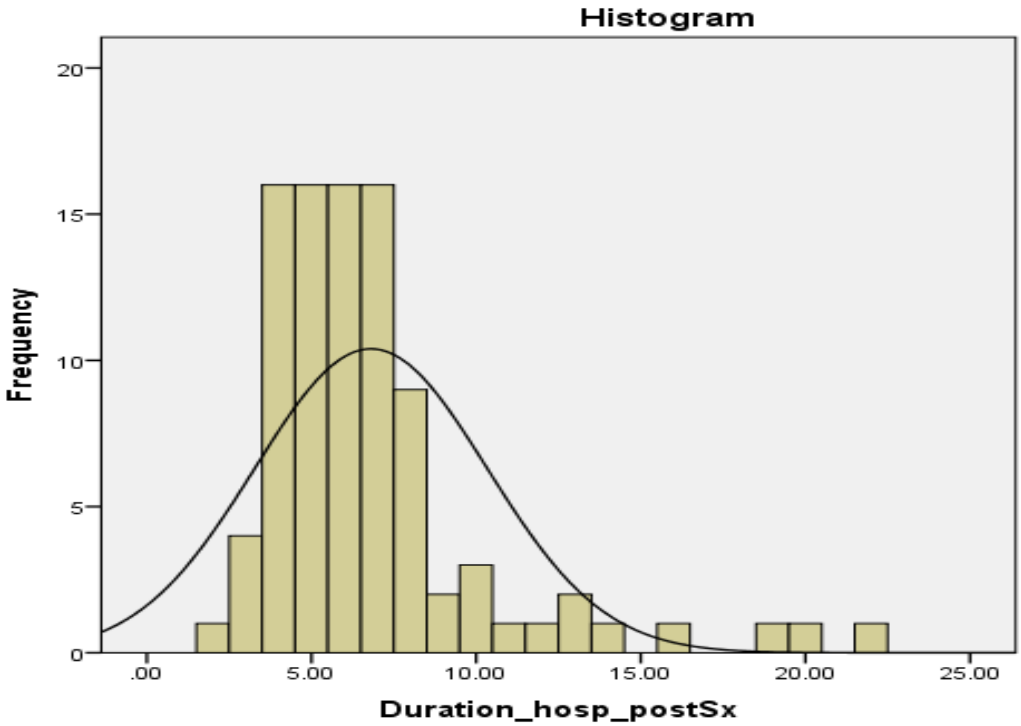
-



The mean duration of the presentation was 6.8 (+-3.5 months). The minimum duration noted in our study was 2 months and the maximum duration of symptoms was 22 months.

- **The average duration of hospital stays following surgery:**

The average duration of hospital stay following surgery was 5-7 days. The mean duration of hospital stay was 6.8 days. Graphical presentation of hospital stay has been shown below.



- **On clinical examination**

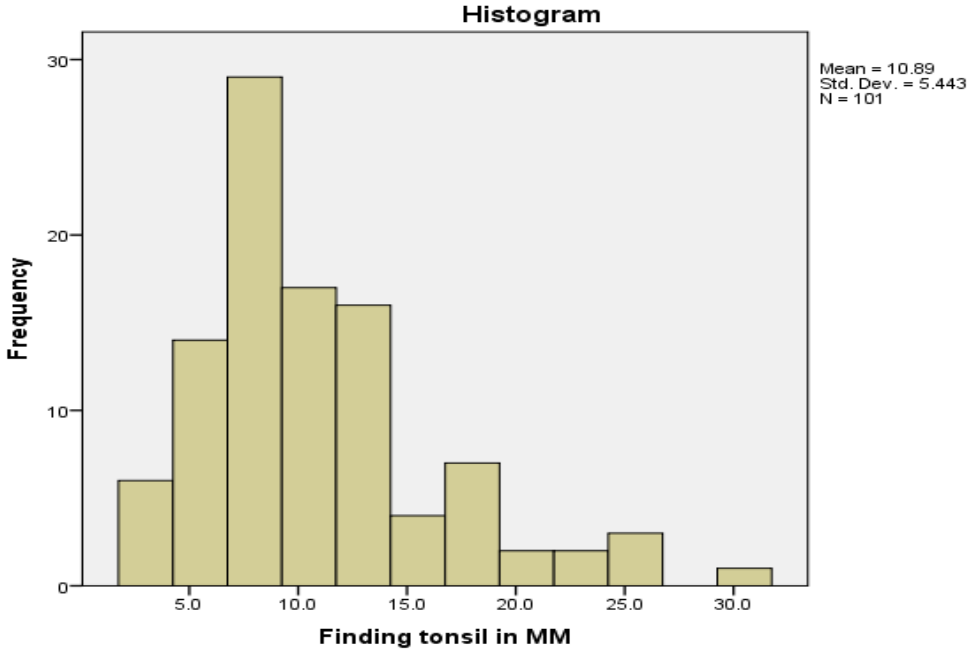
- **Cranial nerve involvement:**

On clinical examination, we noticed 5th cranial nerve was most commonly involved (N 14, 13.9%) followed by the lower cranial nerve 9th-10th (12 and 16 respectively). The least commonly involved nerve was the 12th cranial nerve (N 1)

- **Nystagmus** (upbeat-downbeat, gaze-evoked, horizontal) was seen in almost 50 patients which comprise 49.5% of the study population.
- Hand wasting was seen in 27 patients with grip weakness in 10 patients and limb weakness in 16 patients. a cerebellar sign was seen in 21 patients.

- **Radiological parameters:**

Tonsilar descent was seen in almost all patients. Minimum descent was 3 mm and maximum descent recorded was 30mm. the mean tonsillar descend was 10.88mm (SD 5.4mm)



➢ **Group-wise distribution of tonsilar decent:**

Tonsillar decent	frequency	percentage
<= 3 mm	1	1.0
3.01 - 5 mm	8	7.9
5.01 - 10 mm	52	51.5
10.01 - 15 mm	23	22.8
15.01 - 20 mm	11	10.9
> 20 mm	6	5.9
Total	101	100.0

➢

The majority of patients fell into the 5-15mm tonsillar descent group.

➢ **Radiological relation of the tonsilar tip with foramen magnum:**

	frequency	percentage
Below foramen magnum but above C1	47	46.5
Between C1 and C2	32	31.7
At or below C2	22	21.8
Total	101	100.0

➢

➢ **The dominant side of decent:**

To study the relation of tonsilar descent with scoliosis and syrinx, we studied the dominant site of tonsilar descent. In our study, 32 patients were right side tonsilar descent dominant and 29 patients showed dominant left side tonsilar descent. 40 patients don't show any dominant side of tonsilar descent, in another way both side tonsilar tip were equally herniated from the foramen magnum.

The dominant side of tonsilar decent	frequency	percentage
Same or central	40	39.6
Right	32	31.7
Left	29	28.7
Total	101	100.0

➢ **Preoperative MRI characteristics of syrinx:**

Cervico-dorsal location of syrinx was noted in 51 cases and pure cervical syrinx was seen in 27 cases. Focal syrinx was noted in 2 cases while holocord syrinx was noted in

13 cases. Syrinx was noted as symmetrical or asymmetrical shaped on axial T2WI MRI at the widest part of the syrinx. 16 patients showed postoperative persistent syrinx of the same size, among these 2 patients required re-do surgery and adhesiolysis on follow up due to persistent headache, 2 patients required syringosubarachnoid shunt, 1 patient had progressive scoliosis which was advised deformity correction surgery on follow up. 6 patients showed an increase in the size of syrinx on follow up MRI. Among this 1 patient expired due to respiratory complications and 1 patient had scoliosis progression without significant symptoms.

- **Association between the dominant side of tonsilar decent and syrinx diameter:**

We found if tonsils are equally descended from the foramen magnum, the mean maximum syrinx diameter was 7.5mm with SD of 2.9 and % CI 6.6-8.5. Right side dominant tonsilar decent showed mean maximum diameter of syrinx was 8.2 with SD of 2.8mm. On the left side of the dominant tonsilar decent mean maximum diameter of syrinx was 9.6mm with SD of 2.8mm. On multivariate analysis either left or right side dominant tonsilar decent was significantly associated with the large diameter of syrinx compared to centrally and equally descended tonsils from the foramen magnum.

- **Scoliosis:**

A total of 20 patients were found to have preoperative scoliosis who underwent foramen magnum decompression. The mean diameter of the syrinx in scoliosis was 8.3mm with SD of 3.02. P-value was 0.548 which shows there was no significant relation between syrinx diameter and scoliosis.

- **Improvement:**

We noted 92.07% of patients in our study improved after surgery (p=0.013). We found 2 patients had no changes in syrinx size on follow-up and persistent symptoms which required re-exploration and adhesiolysis along with syringe-subarachnoid shunt and after that they improved.

➢ **Complications:**

Our study demonstrated 2 patients had wound infection and gapping which were managed with OPD bases dressing. 1 patient had pseudomeningocele, but he was asymptomatic and did not require further interventions. One patient with age 15 years, CM with SM underwent FMD, developed post-operative quadriplegia which improved over 2 weeks to the ambulant stage. We could not find any significant factor responsible for the immediate quadriplegia. One patient with preoperative poor respiratory reserve expired following surgery due to respiratory complications.

DISCUSSION

Chiari malformation I (CM I) includes a group of entities of congenital or acquired etiologies which has the common feature of cerebellar tonsil descent below the foramen magnum. (100) Syringomyelia (SM) associated with CM I has been reported 30-70%.(101,102) and 90% SM is associated with obstruction of CSF flow at the foramen magnum level. (103)

Clinical features:

Clinical signs and symptoms in CM are due to compression of brainstem near foramen magnum or due to stretching of dura mater and lower cranial nerve by tonsillar descent or due to syrinx. In our study, the most common presentation was cough headache and the least common presentation was a subjective motor weakness. The mean age of presentation in our study is 29.83 years with female>male. Mehmet Sabri et al and zhang et al have reported similar findings in their studies.(104,105)

We found 31 (31.69%) patients with muscle weakness in form of decreased power or grip weakness on examination. for analysis, we have combined the muscle weakness as symptoms and clinically demonstrated weakness. This is due to some patients who may report mere atrophy or altered sensation as a weakness. Several studies have reported muscle weakness as an insignificant presentation. This weakness can be a significant finding that occurred in 60% of Dyste et al and 80% of Gonçalves da Silva et al series.(106,107)

We found 26.73 % of patients had gross visible muscular atrophy involving the hand or upper limb. In the Dyste et al series 32% of patients were reported with atrophy and milhorat et al reported 10.5% of patients with atrophy in CM with SM and 0.7% in CM without SM.(40,106)

In our study, the most common sensory finding was decreased generalized sensation in 39% of the total patient followed by 14.8% suspended sensory loss and 15.8 % dissociative sensory loss. Trigeminal nerve involvement in form of sensory disturbance

over the face was the most common finding followed by lower cranial nerve involvement. The overall sensory disturbance was 70.6%. Arruda et al reported 86.6% thermogenic dissociation which is quite similar to our study. (102)
Above mentioned findings atrophy or wasting of upper limb muscle, motor weakness, and sensory findings are attributed to both CM and SM. (31,108)

Posterior column signs and symptoms include proprioception impairment, positive Romberg sign, cotton wool sensation, and paraesthesia. We found 21 patients with posterior column involvement. All these patients had tonsillar ectopia ranging between 9-14mm and syrinx diameter >10mm. This explains posterior column signs and symptoms are due to brainstem compression by tonsillar ectopia and posterior column involvement by large syrinx. Similar findings were reported in Dyste et al and Bindall et al. In 60% of Dyste et al. cases 20% were due to brainstem compression. In Bindal et al, 83.3% of cases of brainstem compression combined with SM, and in 100% of cases with only SM symptomatology. (105,106,109) Studies have reported posterior column symptoms significantly associated with a large syrinx. (109)

Diagnosis: in our study 59.4% of a patient diagnosed within 1 year of onset of symptoms. Mehmet Sabri et al reported 64% of patients diagnosed within 36 months of the onset of symptoms. This can be attributed to well-educated people in Kerala and the best healthcare facility. (15,110)

The factor contributing to improvement/failure:

The postoperative analysis showed there was a significant improvement in a patient with CM I following foramen magnum decompression. The most common presentation was cough headache which improved significantly following foramen magnum decompression. We noted 92.07% of patients in our study improved after surgery ($p=0.013$). We found 2 patients had no changes in syrinx size on follow-up and persistent symptoms which required re-exploration and adhesiolysis along with

syringe-subarachnoid shunt and after that they improved. One patient with age 15 years, CM with SM underwent FMD, developed post-operative quadriplegia which improved over 2 weeks to the ambulant stage. We could not find any significant factor responsible for the immediate quadriplegia. One patient with preoperative poor respiratory reserve expired following surgery due to respiratory complications. We noticed all the above patients had poor or insufficient improvement in CSF flow across the foramen magnum. The probable mechanism for symptomatic improvement was the restoration of CSF flow across the foramen magnum. We didn't find any significant correlation between age, sex, and duration of symptoms with postoperative improvement in symptoms.

Recently, goel has proposed fixation of the atlanto-axial joint considering subtle instability at this joint causing symptomatology. (21,97,111) However, there are many critics regarding this management. We strongly condemn the fixation of the C1-C2 joint instead of the FMD.

Foramen magnum decompression surgery aims to relieve pressure and provide space to flow CSF across the foramen magnum which relieves the patient's symptoms. It also prevents further progression of SM and prevents potential damage to neural tissue. Many studies have reported 80% improvement in symptoms and 15% without progression of symptoms. few patients deteriorate even after good surgery may be due to the formation of adhesions and failure to establish CSF flow across the foramen magnum. (15,110,112–114) In our institute, we do foramen magnum decompression with C1 laminectomy and Lax duroplasty unless it is contraindicated. Some authors have claimed simple foramen magnum decompression without laminectomy may result in acute Cervico-dural angle indicating residual compression. (115)

Changes in syrinx:

In our study, we noticed the patient who did not show any change in syrinx size following surgery or who demonstrate an increase in the size of syrinx were presented late after the onset of symptoms. We noted most patients presented between 24-48 months after the onset of symptoms. Bogdanov et al reported persistent symptoms even after resolution of syringomyelia. It has been noted in autopsy long-standing syringomyelia provokes gliosis formation inside the cord which leads to progressive neurological loss even after successful surgery and collapse of syringomyelia. Whatever is the mechanism young patient and family must be counseled properly before surgery. (116,117) if FMD fails other possible causes should be kept in mind, for example, hydrocephalus, insufficient FMD, and spinal instability should be evaluated and treated. (118)

Nakamura et al noted the morphology of the syrinx at the spinal cord level coincided with the dermatome at the site of pain and reported persistent post-operative pain seen in the deviated type of syrinx. in another way posterolateral located syrinx coincides with the dorsal horn of the gray matter of the spinal cord, which is responsible for irreversible changes. Milhorat et al also reported persistent intractable pain after surgery is due to extent of syrinx to the dorsal horn of the spinal cord. (119,120) These should be kept in mind while treating CM with SM. In our study due to retrospective in nature, we couldn't measure pain score. We shall consider a prospective study to overcome the above limitation.

Milhorat and colleagues observed that a reduction in the size of syrinx following surgery is usually seen in the central type of syrinx. Central types of syrinx are due to enlargement of the central canal and they reverse following improvement of CSF flow. They noted that nerve fibers in the afferent pathway that passes the anterior gray commissure are likely to undergo functional improvement and subsequent amelioration of differential pain due to reduction of the syrinx size resultant decompressive effect. (121) in our study, we have noticed in a few cases eccentrically placed and irregular-shaped syrinx may not resolve completely. We also noticed asymmetrical tonsillar descent is significantly associated with larger syrinx diameter.

Mortality and morbidity:

We have one mortality in our study that was not related to the surgical procedures. That patient had larger syrinx and preoperative poor respiratory reserve which may have aggravated after surgery leading to aspiration pneumonia and worsening of lung condition.

LIMITATIONS OF THE STUDY

- A major limitation of our study is retrospective design.

- The data and their interpretation are limited by completeness of medical records and consistency between preoperative evaluations and availability of standing X-ray film.

- A study of scoliosis was done using available X-ray film and MRI film and only coronal parameters have been evaluated. A further prospective study using proper X-ray standing film with a coronal and sagittal view is required.

- Being a tertiary care center what cases are referred to us are only included in our study which may not represent the actual population.

- We have excluded complex Chiari malformation cases. Future study is required including all types of cases.

- A detailed study including preoperative and postoperative CSF flow study is required for the understanding of the exact mechanism responsible for the resolution of symptoms and syrinx following surgery.

CONCLUSION

- Foramen magnum decompression with C1 arch excision and lax Duroplast is a safe and effective procedure for Chiari malformation with or without syringomyelia.

- Larger syrinx and long duration of symptoms may affect syrinx resolution, which should be kept in mind during patient counseling.

- Preoperative good pulmonary condition and atlanto-axial should be ruled out as these may flip the usual outcome of simple surgery.

- We recommend atlanto-axial fixation only if clear instability is demonstrated on dynamic X-ray. We recommend dynamic X-rays in all cases which are planned for FMD.

- Syringo-subarachnoid shunt is a simple and effective measure of management for persistent large syrinx if CSF flow presents across the foramen magnum on flow study.

REFERENCE

1. Barkovich AJ, Wippold FJ, Sherman JL, Citrin CM. Significance of cerebellar tonsillar position on MR. AJNR Am J Neuroradiol. 1986 Oct;7(5):795–9.

2. Neurologic syndromes accompanying developmental anomalies of occipital bone, atlas, and axis. Arch Neurol Psychiatry. 1941;

3. Chorobski J, Stepien L. On the syndrome of Arnold-Chiari; report of a case. J Neurosurg. 1948 Sep;5(5):495–500.

4. Chiari H. Ueber Veränderungen des Kleinhirns infolge von Hydrocephalie des Grosshirns1). DMW - Dtsch Med Wochenschr. 1891 Oct;17(42):1172–5.

5. The surgical treatment of Arnold-Chiari malformation in adults; an explanation of its mechanism and importance of encephalography in diagnosis - PubMed [Internet]. [cited 2020 Oct 8]. Available from: https://pubmed.ncbi.nlm.nih.gov/15415776/

6. Koehler PJ. Chiari's description of cerebellar ectopy (1891). With a summary of Cleland's and Arnold's contributions and some early observations on neural-tube defects. J Neurosurg. 1991 Nov;75(5):823–6.

7. McConnell: A deformity of the hind-brain associated... - Google Scholar [Internet]. [cited 2020 Oct 8]. Available from: https://scholar.google.com/scholar_lookup?title=Deformity%20of%20hind-brain%20associated%20with%20internal%20hydrocephalus%3B%20its%20relation%20to%20Arnold-Chiari%20malformation&journal=Brain&volume=61&pages=415-429&publication_year=1938&author=McConnell%2CA.%20A.&author=Parker%2CH.%20L.

8. Graftdijk: Over hydrocephalus - Google Scholar [Internet]. [cited 2020 Oct 8]. Available from: https://scholar.google.com/scholar_lookup?title=Over%20hydrocephalus&author=CJ.%20Houweninge%20Graftdijk&publication_year=1932

9. THE MECHANISM OF INTERNAL HYDROCEPHALUS IN SPINA BIFIDA | Brain | Oxford Academic [Internet]. [cited 2020 Oct 8]. Available from: https://academic.oup.com/brain/article-abstract/58/2/203/319845?redirectedFrom=fulltext

10. Steele GH. The Arnold-Chiari malformation. Br J Surg. 1947 Jan;34(135):280–2.

11. Padget DH. Development of so-called dysraphism; with embryologic evidence of clinical Arnold-Chiari and Dandy-Walker malformations. Johns Hopkins Med J. 1972 Mar;130(3):127–65.

12. Ingraham FD, Scott HW. Spina Bifida and Cranium Bifidum: The Arnold-Chiari Malformation: a Study of 20 Cases. N Engl J Med. 1943 Jul 15;229(3):108–14.

13. Schady W, Metcalfe RA, Butler P. The incidence of craniocervical bony anomalies in the adult Chiari malformation. J Neurol Sci. 1987 Dec;82(1–3):193–203.

14. Smoker WR. Craniovertebral junction: normal anatomy, craniometry, and congenital anomalies. RadioGraphics. 1994 Mar 1;14(2):255–77.

15. Tubbs RS, Beckman J, Naftel RP, Chern JJ, Wellons JC, Rozzelle CJ, et al. Institutional experience with 500 cases of surgically treated pediatric Chiari malformation Type I: Clinical article. J Neurosurg Pediatr. 2011 Mar 1;7(3):248–56.

16. Gardner WJ. Hydrodynamic factors in Dandy-Walker and Arnold-Chiari malformations. Childs Brain. 1977;3(4):200–12.

17. Masters CL. Pathogenesis of the Arnold-Chiari Malformation: The Significance of Hydrocephalus and Aqueduct Stenosis. J Neuropathol Exp Neurol. 1978 Jan 1;37(1):56–74.

18. Peach B. The Arnold-Chiari Malformation: Morphogenesis. Arch Neurol. 1965 May 1;12(5):527.

19. Lichtenstein BW. DISTANT NEUROANATOMIC COMPLICATIONS OF SPINA BIFIDA (SPINAL DYSRAPHISM): HYDROCEPHALUS, ARNOLD-CHIARI DEFORMITY, STENOSIS OF THE AQUEDUCT OF SYLVIUS, ETC.; PATHOGENESIS AND PATHOLOGY. Arch Neurol Psychiatry. 1942 Feb 1;47(2):195.

20. Lemire RJ. The graphic states: From syringomyelia to anencephaly. W. J. Gardner. Excerpta Medica, Amsterdam. 201 pp. 1973. Teratology. 1974;9(2):248–9.

21. Goel A. Is Chiari malformation nature's protective "air-bag"? Is its presence diagnostic of atlantoaxial instability? J Craniovertebral Junction Spine. 2014 Jul;5(3):107–9.

22. Bell JE, Gordon A, Maloney AFJ. The Association of Hydrocephalus and Arnold-Chiari Malformation with Spina Bifida in the Fetus. Neuropathol Appl Neurobiol. 1980;6(1):29–39.

23. Cameron AH. The Arnold-Chiari and other neuro-anatomical malformations associated with spina bifida. J Pathol Bacteriol. 1957;73(1):195–211.

24. Gardner WJ, Abdullah AF, McCormack LJ. The Varying Expressions of Embryonal Atresia of the Fourth Ventricle in Adults: Arnold-Chiari

Malformation, Dandy-Walker Syndrome, “Arachnoid” Cyst of the Cerebellum, and Syringomyelia. J Neurosurg. 1957 Nov 1;14(6):591–607.

25. Marin-Padilla M, Marin-Padilla TM. Morphogenesis of experimentally induced Arnold-Chiari malformation. J Neurol Sci. 1981 Apr;50(1):29–55.

26. Barry A, Patten BM, Stewart BH. Possible Factors in the Development of the Arnold-Chiari Malformation. J Neurosurg. 1957 May;14(3):285–301.

27. ARNOLD-CHIARI MALFORMATION AND ITS OPERATIVE TREATMENT | Archives of Neurology & Psychiatry | JAMA Network [Internet]. [cited 2020 Oct 8]. Available from: https://jamanetwork.com/journals/archneurpsyc/article-abstract/647726

28. Daniel PM, Strich SJ. Some Observations on the Congenital Deformity of the Central Nervous System Known as the Arnold-Chiari Malformation. J Neuropathol Exp Neurol. 1958 Apr 1;17(2):255–66.

29. McLone DG, Knepper PA. The Cause of Chiari II Malformation: A Unified Theory. Pediatr Neurosurg. 1989;15(1):1–12.

30. Roth M. Cranio-cervical growth collision: another explanation of the Arnold-Chiari malformation and basilar impression. Neuroradiology. 1986 May 1;28(3):187–94.

31. Williams B. On the pathogenesis of syringomyelia: a review. J R Soc Med. 1980 Nov;73(11):798–806.

32. Oldfield EH, Muraszko K, Shawker TH, Patronas NJ. Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils: Implications for diagnosis and treatment. J Neurosurg. 1994 Jan 1;80(1):3–15.

33. Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, Wolpert C, et al. Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. Neurosurgery. 1999 May;44(5):1005–17.

34. Turgut M. Chiari type I malformation in two monozygotic twins. Br J Neurosurg. 2001 Jan 1;15(3):279–80.

35. Atkinson JLD, Kokmen E, Miller GM. Evidence of Posterior Fossa Hypoplasia in the Familial Variant of Adult Chiari I Malformation: Case Report. Neurosurgery. 1998 Feb 1;42(2):401–4.

36. Ellertsson ÁB. Syringomyelia and Other Cystic Spinal Cord Lesions. Acta Neurol Scand. 1969;45(4):403–17.

37. Gardner WJ, Angel J. The cause of syringomyelia and its surgical treatment. Cleve Clin Q. 1958 Jan;25(1):4–8.

38. Heiss JD, Patronas N, DeVroom HL, Shawker T, Ennis R, Kammerer W, et al. Elucidating the pathophysiology of syringomyelia. J Neurosurg. 1999 Oct 1;91(4):553–62.

39. Klekamp J. The Pathophysiology of Syringomyelia – Historical Overview and Current Concept. Acta Neurochir (Wien). 2002 Jul 1;144(7):649–64.

40. Milhorat TH, Capocelli AL, Anzil AP, Kotzen RM, Milhorat RH. Pathological basis of spinal cord cavitation in syringomyelia: analysis of 105 autopsy cases. J Neurosurg. 1995 May 1;82(5):802–12.

41. Milhorat TH, Capocelli AL, Kotzen RM, Bolognese P, Heger IM, Cottrell JE. Intramedullary Pressure in Syringomyelia: Clinical and Pathophysiological Correlates of Syrinx Distension. Neurosurgery. 1997 Nov 1;41(5):1102–10.

42. Milhorat TH. Classification of syringomyelia. Neurosurg Focus. 2000 Mar 1;8(3):1–6.

43. Chern JJ, Gordon AJ, Mortazavi MM, Tubbs RS, Oakes WJ. Pediatric Chiari malformation Type 0: a 12-year institutional experience: Clinical article. J Neurosurg Pediatr. 2011 Jul 1;8(1):1–5.

44. Williams B. The distending force in the production of "communicating syringomyelia." Lancet Lond Engl. 1969 Jul 26;2(7613):189–93.

45. Harrison DK. Pathology of Syringomyelia Due to Chiari Malformations. In: Tubbs RS, Oakes WJ, editors. The Chiari Malformations [Internet]. New York, NY: Springer; 2013 [cited 2020 Oct 8]. p. 121–5. Available from: https://doi.org/10.1007/978-1-4614-6369-6_9

46. Rusbridge C, Greitz D, Iskandar BJ. Syringomyelia: Current Concepts in Pathogenesis, Diagnosis, and Treatment. J Vet Intern Med. 2006;20(3):469–79.

47. Syringomyelia - Diagnosis, and Treatment | Jörg Klekamp | Springer [Internet]. [cited 2020 Oct 8]. Available from: https://www.springer.com/gp/book/9783642626517

48. Gardner WJ, Angel J. The Mechanism of Syringomyelia and Its Surgical Correction. Neurosurgery. 1959 Jan 1;6(CN_suppl_1):131–40.

49. The pathogenesis of syringomyelia associated with lesions at the foramen magnum: a critical review of existing theories and proposal of a new hypothesis - PubMed [Internet]. [cited 2020 Oct 8]. Available from: https://pubmed.ncbi.nlm.nih.gov/15140600/

50. Enzmann DR, Pelc NJ. Normal flow patterns of intracranial and spinal cerebrospinal fluid defined with phase-contrast cine MR imaging. Radiology. 1991 Feb;178(2):467–74.

51. Greitz D. Cerebrospinal fluid circulation and associated intracranial dynamics. A radiologic investigation using MR imaging and radionuclide cisternography. Acta Radiol Suppl. 1993;386:1–23.

52. Hofmann E, Warmuth-Metz M, Bendszus M, Solymosi L. Phase-contrast MR imaging of the cervical CSF and spinal cord: volumetric motion analysis in patients with Chiari I malformation. AJNR Am J Neuroradiol. 2000 Jan;21(1):151–8.

53. Greitz D. Radiological assessment of hydrocephalus: new theories and implications for therapy. Neurosurg Rev. 2004 Jul;27(3):145–65; discussion 166-167.

54. Miller E, Widjaja E, Blaser S, Dennis M, Raybaud C. The old and the new: supratentorial MR findings in Chiari II malformation. Childs Nerv Syst. 2008 May 1;24(5):563–75.

55. Andica C, Soetikno RD. Chiari malformation type III: Case report and review of the literature. Radiol Case Rep [Internet]. 2015 Nov 6 [cited 2020 Oct 8];8(3). Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4900115/

56. Mikulis DJ, Diaz O, Eglin TK, Sanchez R. Variance of the position of the cerebellar tonsils with age: a preliminary report. Radiology. 1992 Jun 1;183(3):725–8.

57. Jatavallabhula NS, Armstrong J, Sgouros S, Whitehouse W. Spontaneous resolution of isolated Chiari I malformation. Childs Nerv Syst ChNS Off J Int Soc Pediatr Neurosurg. 2006 Feb;22(2):201–3.

58. Miller JH, Limbrick DD, Callen M, Smyth MD. Spontaneous resolution of Chiari malformation Type I in monozygotic twins. J Neurosurg Pediatr. 2008 Nov;2(5):317–9.

59. Spontaneous Resolution of Chiari I Malformation and Syringomyelia: Case Report and Review of the Literature | Neurosurgery | Oxford Academic [Internet]. [cited 2020 Nov 30]. Available from: https://academic.oup.com/neurosurgery/article/48/3/664/2751746

60. Williams B. Cough Headache due to Craniospinal Pressure Dissociation. Arch Neurol. 1980 Apr 1;37(4):226–30.

61. Pascual J, Oterino A, Berciano J. Headache in type I Chiari malformation. Neurology. 1992 Aug 1;42(8):1519–1519.

62. Kowal L, Yahalom C, Shuey NH. Chiari 1 malformation presenting as strabismus. Binocular Vis Strabismus Q. 2006;21(1):18–26.

63. Tisell M, Wallskog J, Linde M. Long-term outcome after surgery for Chiari I malformation. Acta Neurol Scand. 2009;120(5):295–9.

64. Massimi L, Della Pepa GM, Caldarelli M, Di Rocco C. Abrupt clinical onset of Chiari type I/syringomyelia complex: clinical and physiopathological implications. Neurosurg Rev. 2012 Jul 1;35(3):321–9.

65. Yarbrough CK, Powers AK, Park TS, Leonard JR, Limbrick DD, Smyth MD. Patients with Chiari malformation Type I presenting with acute neurological deficits: case series. J Neurosurg Pediatr. 2011 Mar;7(3):244–7.

66. Ziegler DK, Mallonee W. Chiari-1 malformation, migraine, and sudden death. Headache. 1999 Jan;39(1):38–41.

67. Goodwin D, Halvorson AR. Chiari I malformation presenting as downbeat nystagmus: clinical presentation, diagnosis, and management. Optom St Louis Mo. 2012 Feb 15;83(2):80–6.

68. Shaikh AG, Ghasia FF. Neuro-ophthalmology of type 1 Chiari malformation. Expert Rev Ophthalmol. 2015;10(4):351–7.

69. Paul KS, Lye RH, Strang FA, Dutton J. Arnold-Chiari malformation. Review of 71 cases. J Neurosurg. 1983 Feb;58(2):183–7.

70. Waqar M, Vohra AH. Dissociated sensory loss and muscle wasting in a young male with headaches: syringomyelia with type 1 Arnold-Chiari malformation. BMJ Case Rep. 2013 Oct 29;2013.

71. Scelsa SN. Syringomyelia presenting as ulnar neuropathy at the elbow. Clin Neurophysiol Off J Int Fed Clin Neurophysiol. 2000 Sep;111(9):1527–30.

72. Banik R, Lin D, Miller NR. Prevalence of Chiari I malformation and cerebellar ectopia in patients with pseudotumor cerebri. J Neurol Sci. 2006 Aug 15;247(1):71–5.

73. Sinclair N, Assaad N, Johnston I. Pseudotumour cerebri occurring in association with the Chiari malformation. J Clin Neurosci Off J Neurosurg Soc Australas. 2002 Jan;9(1):99–101.

74. Johnston I, Jacobson E, Besser M. The acquired Chiari malformation and syringomyelia following spinal CSF drainage: a study of incidence and management. Acta Neurochir (Wien). 1998;140(5):417–27; discussion 427-428.

75. Wang J, Alotaibi NM, Samuel N, Ibrahim GM, Fallah A, Cusimano MD. Acquired Chiari Malformation and Syringomyelia Secondary to Space-Occupying Lesions: A Systematic Review. World Neurosurg. 2017 Feb;98:800-808.e2.

76. Rahman M, Bidari SS, Quisling RG, Friedman WA. Spontaneous intracranial hypotension: dilemmas in diagnosis. Neurosurgery. 2011 Jul;69(1):4–14; discussion 14.

77. Samii C, Möbius E, Weber W, Heienbrok HW, Berlit P. Pseudo Chiari type I malformation secondary to cerebrospinal fluid leakage. J Neurol. 1999 Mar;246(3):162–4.

78. Klekamp J. Chiari I malformation with and without basilar invagination: a comparative study. Neurosurg Focus. 2015 Apr 1;38(4):E12.

79. Goel A, Bhatjiwale M, Desai K. Basilar invagination: a study based on 190 surgically treated patients. J Neurosurg. 1998 Jun;88(6):962–8.

80. Förander P, Sjåvik K, Solheim O, Riphagen I, Gulati S, Salvesen Ø, et al. The case for duraplasty in adults undergoing posterior fossa decompression for Chiari I malformation: a systematic review and meta-analysis of observational studies. Clin Neurol Neurosurg. 2014 Oct;125:58–64.

81. Nohria V, Oakes J. Chiari I Malformation: A Review of 43 Patients. Pediatr Neurosurg. 1990;16(4–5):222–7.

82. Ellenbogen RG, Armonda RA, Shaw DW, Winn HR. Toward a rational treatment of Chiari I malformation and syringomyelia. Neurosurg Focus. 2000 Mar 15;8(3):E6.

83. Deng X, Wu L, Yang C, Tong X, Xu Y. Surgical treatment of Chiari I malformation with ventricular dilation. Neurol Med Chir (Tokyo). 2013;53(12):847–52.

84. Hayhurst C, Osman-Farah J, Das K, Mallucci C. Initial management of hydrocephalus associated with Chiari malformation Type I-syringomyelia complex via endoscopic third ventriculostomy: an outcome analysis. J Neurosurg. 2008 Jun;108(6):1211–4.

85. Royo-Salvador MB, Solé-Llenas J, Doménech JM, González-Adrio R. Results of the section of the filum terminale in 20 patients with syringomyelia, scoliosis and Chiari malformation. Acta Neurochir (Wien). 2005 May;147(5):515–23; discussion 523.

86. Milhorat TH, Bolognese PA, Nishikawa M, Francomano CA, McDonnell NB, Roonprapunt C, et al. Association of Chiari malformation type I and tethered cord syndrome: preliminary results of sectioning filum terminale. Surg Neurol. 2009 Jul;72(1):20–35.

87. Fenoy AJ, Menezes AH, Fenoy KA. Craniocervical junction fusions in patients with hindbrain herniation and syringohydromyelia. J Neurosurg Spine. 2008 Jul;9(1):1–9.

88. Kohno K, Sakaki S, Shiraishi T, Matsuoka K, Okamura H. Successful treatment of adult Arnold-Chiari malformation associated with basilar impression and syringomyelia by the transoral anterior approach. Surg Neurol. 1990 Apr;33(4):284–7.

89. Strahle J, Muraszko KM, Buchman SR, Kapurch J, Garton HJL, Maher CO. Chiari malformation associated with craniosynostosis. Neurosurg Focus. 2011 Sep;31(3):E2.

90. Durham SR, Fjeld-Olenec K. Comparison of posterior fossa decompression with and without duraplasty for the surgical treatment of Chiari malformation Type I in pediatric patients: a meta-analysis. J Neurosurg Pediatr. 2008 Jul;2(1):42–9.

91. Zhao J-L, Li M-H, Wang C-L, Meng W. A Systematic Review of Chiari I Malformation: Techniques and Outcomes. World Neurosurg. 2016 Apr;88:7–14.

92. Xu H, Chu L, He R, Ge C, Lei T. Posterior fossa decompression with and without duraplasty for the treatment of Chiari malformation type I-a systematic review and meta-analysis. Neurosurg Rev. 2017 Apr;40(2):213–21.

93. Shamji MF, Ventureyra ECG, Baronia B, Nzau M, Vassilyadi M. Classification of symptomatic Chiari I malformation to guide surgical strategy. Can J Neurol Sci J Can Sci Neurol. 2010 Jul;37(4):482–7.

94. Yeh DD, Koch B, Crone KR. Intraoperative ultrasonography used to determine the extent of surgery necessary during posterior fossa decompression in children with Chiari malformation type I. J Neurosurg. 2006 Jul;105(1 Suppl):26–32.

95. Behari S, Kalra SK, Kiran Kumar MV, Salunke P, Jaiswal AK, Jain VK. Chiari I malformation associated with atlanto-axial dislocation: focussing on the anterior cervico-medullary compression. Acta Neurochir (Wien). 2007 Jan 1;149(1):41–50.

96. Menezes AH. Primary craniovertebral anomalies and the hindbrain herniation syndrome (Chiari I): database analysis. Pediatr Neurosurg. 1995;23(5):260–9.

97. Goel A. Is atlantoaxial instability the cause of Chiari malformation? Outcome analysis of 65 patients treated by atlantoaxial fixation. J Neurosurg Spine. 2015 Feb 1;22(2):116–27.

98. Jea A. Editorial: Chiari malformation I surgically treated with atlantoaxial fixation. J Neurosurg Spine. 2015 Feb 1;22(2):113–5.

99. Brockmeyer DL, Oakes WJ, Rozzelle C, Johnston J, Rocque BG, Anderson RCE, et al. Letter to the Editor: Chiari malformation Type 1 and atlantoaxial

instability: a letter from the Pediatric Craniocervical Society. J Neurosurg Spine. 2015 Dec;23(6):820–3.

100. Considerations in the diagnosis and treatment of syringomyelia and the Chiari malformation - PubMed [Internet]. [cited 2020 Nov 29]. Available from: https://pubmed.ncbi.nlm.nih.gov/7086497/

101. Surgery for Syringomyelia: An Analysis Based on 163 Surgical Cases | SpringerLink [Internet]. [cited 2020 Nov 29]. Available from: https://link.springer.com/article/10.1007/s007010050038

102. Arruda JAM de, Costa CM de C, Tella Jr OI de. Results of the treatment of syringomyelia associated with Chiari malformation: analysis of 60 cases. Arq Neuropsiquiatr. 2004 Jun;62(2a):237–44.

103. McGirt MJ, Atiba A, Attenello FJ, Wasserman BA, Datoo G, Gathinji M, et al. Correlation of hindbrain CSF flow and outcome after surgical decompression for Chiari I malformation. Childs Nerv Syst. 2008 Jul;24(7):833–40.

104. Zhang ZQ, Chen YQ, Chen YA, Wu X, Wang YB, Li XG. Chiari I malformation associated with syringomyelia: a retrospective study of 316 surgically treated patients. Spinal Cord. 2008 May;46(5):358–63.

105. Godzik J, Kelly MP, Radmanesh A, Kim D, Holekamp TF, Smyth MD, et al. Relationship of syrinx size and tonsillar descent to spinal deformity in Chiari malformation Type I with associated syringomyelia: Clinical article. J Neurosurg Pediatr. 2014 Apr;13(4):368–74.

106. Dyste GN, Menezes AH, VanGilder JC. Symptomatic Chiari malformations. An analysis of presentation, management, and long-term outcome. J Neurosurg. 1989 Aug;71(2):159–68.

107. DA S, Taricco M, FARIAS B, Neves V, LUCENA F. Aracnoidite constritiva causada por pantopaque resultando em siringomielia e paraparesia: relato de caso. Arq Neuropsiquiatr. 2001 Sep 1;59.

108. Memarpour R, Tashtoush B, Issac L, Gonzalez-Ibarra F. Syringomyelia with Chiari I Malformation Presenting as Hip Charcot Arthropathy: A Case Report and Literature Review. Case Rep Neurol Med [Internet]. 2015 [cited 2020 Nov 30];2015. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4322850/

109. Bindal AK, Dunsker SB, Tew JM. Chiari I Malformation. Neurosurgery. 1995 Dec 1;37(6):1069–74.

110. Di X. Endoscopic Suboccipital Decompression on Pediatric Chiari Type I. Minim Invasive Neurosurg MIN. 2009 Jul 1;52:119–25.

111. Can foramen magnum decompression surgery become historical? - Abstract - Europe PMC [Internet]. [cited 2020 Nov 30]. Available from: http://europepmc.org/article/PMC/4426520

112. Litvack ZN, Lindsay RA, Selden NR. Dura Splitting Decompression for Chiari I Malformation in Pediatric Patients: Clinical Outcomes, Healthcare Costs, and Resource Utilization. Neurosurgery. 2013 Jun 1;72(6):922–9.

113. Kennedy BC, Kelly KM, Phan MQ, Bruce SS, McDowell MM, Anderson RCE, et al. Outcomes after suboccipital decompression without dural opening in children with Chiari malformation Type I. J Neurosurg Pediatr. 2015 Aug;16(2):150–8.

114. Gallo P, Sokol D, Kaliaperumal C, Kandasamy J. Comparison of Three Different Cranio-Cervical Decompression Procedures in Children with Chiari Malformation Type I: Does the Surgical Technique Matter? Pediatr Neurosurg. 2017;52(5):289–97.

115. Importance of C1 laminectomy in foramen magnum decompression surgery: A technical note. - Abstract - Europe PMC [Internet]. [cited 2020 Nov 30]. Available from: http://europepmc.org/article/PMC/4323971

116. Bogdanov EI, Heiss JD, Mendelevich EG. The post-syrinx syndrome: stable central myelopathy and collapsed or absent syrinx. J Neurol. 2006 Jun;253(6):707–13.

117. Shetty J, Kandasamy J, Sokol D, Gallo P. Clinical deterioration despite syringomyelia resolution after successful foramen magnum decompression for Chiari malformation - Case series. Eur J Paediatr Neurol EJPN Off J Eur Paediatr Neurol Soc. 2019 Mar;23(2):333–7.

118. Soleman J, Bartoli A, Korn A, Constantini S, Roth J. Treatment failure of syringomyelia associated with Chiari I malformation following foramen magnum decompression: how should we proceed? Neurosurg Rev. 2019 Sep 1;42.

119. Nakamura M, Chiba K, Nishizawa T, Maruiwa H, Matsumoto M, Toyama Y. Retrospective study of surgery-related outcomes in patients with syringomyelia associated with Chiari I malformation: Clinical significance of changes in the size and localization of syrinx on pain relief. J Neurosurg. 2004 Apr 1;100:241–4.

120. Milhorat TH, Mu HT, LaMotte CC, Milhorat AT. Distribution of substance P in the spinal cord of patients with syringomyelia. J Neurosurg. 1996 Jun;84(6):992–8.

121. Dysesthetic Pain in Patients with Syringomyelia | Neurosurgery | Oxford Academic [Internet]. [cited 2020 Nov 30]. Available from:

https://academic.oup.com/neurosurgery/article-abstract/38/5/940/2812821?redirectedFrom=fulltext

122. Arai T, Ohno K, Takada Y, Aoyagi M, Hirakawa K. Neonatal craniopharyngioma and inference of tumor inception time: case report and review of the literature. Surg Neurol. 2003;60(3):254–9.

123. Arai K, Sato N, Aoki J, Yagi A, Taketomi-Takahashi A, Morita H, et al. MR signal of the solid portion of pilocytic astrocytoma on T2-weighted images: is it useful for differentiation from medulloblastoma? Neuroradiology. 2006;48(4):233–7.

124. Arai K, Sato N, Aoki J, Yagi A, Taketomi-Takahashi A, Morita H, et al. MR signal of the solid portion of pilocytic astrocytoma on T2-weighted images: is it useful for differentiation from medulloblastoma? Neuroradiology. 2006;48(4):233–7.

ANNEXURES

Sree Chitra Tirunal Institute for Medical Sciences & Technology

Proforma

Clinical and radiological outcomes following foramen magnum decompression in patients with chiari malformation

Annexure 1:

History:

Hospital number
Age
Sex
Primary complaint
Age at onset
Evidence of maturation
Presence of back pain
Neurological symptoms, including gait abnormalities, weakness or sensory changes
Feelings about overall appearance and back shape Family history

Clinical examination:

Positive clinical finding
Neck height
Height measurement
Gait check
Foot shape
Skin inspection
Assessment of pubertal development
Neurological examination, including motor, sensory and reflex tests (including abdominal)
The symmetry of shoulders and iliac crest Forward bending test
Adam's forward bend test

Radiological data

Degree of cerebellar tonsil decent(CTD grade) in midsagittal image
Coronal image distance of each side tonsil from foramen magnum margin
Dominant side decent(right/ left)
The ratio of decent (A/B)

Scoliosis measurement:
Cobb angle
Curve direction(left/ right)
Number of vertebrae

Coronal balance in cm

Syrinx length (vertebral level)
Maximum Cord/ syrinx ratio at any level

CSF flow study value

Operative data

Approach
Amount of decompression
Dura opened or not
Duroplasty
Complication

Postoperative scan:

Bony decompression in cm
Amount of C1 arch excision
Positive findings

Annexure 2:

Follow up finding
Positive history
Clinical finding
Bracing type and duration

Radiological data

Degree of cerebellar tonsil decent(CTD grade) in midsagittal image
Coronal image distance of each side tonsil from foramen magnum margin
Dominant side decent(right/ left)
The ratio of decent (A/B)

Scoliosis measurement:
Cobb angle
Curve direction(left/ right)
Number of vertebrae
Coronal balance in cm
Syrinx length (vertebral level)
Maximum Cord/ syrinx ratio at any level

CSF flow study value

SREE CHITRA TIRUNAL INSTITUTE FOR MEDICAL SCIENCES & TECHNOLOGY
THIRUVANANTHAPURAM—695 011, INDIA.
(An Institute of National Importance under Govt.of India)
Phone—(91)0471—2443152 Fax—(91)0471—2446433, 2550728, Grams—Chitramet
email : sct@sctimst.ac.in Web site—www.sctimst.ac.in

CONFIDENTIAL

SREE CHITRA TIRUNAL INSTITUTE FOR MEDICAL SCIENCES AND TECHNOLOGY, TRIVANDRUM
THIRUVANANTHAPURAM-695011.

INSTITUTIONAL ETHICS COMMITTEE (IEC) MEETING
(IEC Regn No. ECR/189/Inst/KL/2013/RR-16)

SCT/IEC/1547 /AUGUST-2020 17.08.2020

Dr. Jaypalsinh Gohil
PDF skullbase neurosurgery
Department of Neurosurgery
SCTIMST, Thiruvananthapuram

Dear Dr. Jayapalsinh Gohil,

The Institutional Ethics Committee reviewed and discussed your application to conduct the study entitled "**CLINICAL AND RADIOLOGICAL OUTCOMES FOLLOWING FORAMEN MAGNUM DECOMPRESSION IN PATIENTS WITH CHIARI MALFORMATION (IEC/1547)**" on July 10-17, 2020.

The following documents were reviewed:

Original documents

1. *Covering Letter addressed to the Chairman, IEC, SCTIMST dated 21.06.2020*	10. *CV of Dr. Jaypalsinh Gohil*
2. *TAC Approval Letter*	11. *CV of Dr Ganesh Divakar*
3. *IEC Application Form*	12. *CV of Dr Krishnakumar K*
4. *Project Proposal*	13. *CV of Dr Mathew Abraham*
5. *Proforma*	14. *CV of Dr Bejoy Thomas*
6. *Patient Information Sheet in English*	15. *CV of Dr Harshavardhan Biradar*
7. *Patient Information Sheet in Malayalam*	16. *Declaration Form*
8. *Informed Consent Form in English*	17. *Covering letter from Dr. Jaypalsinh Gohil dated 26.06.2020*
9. *Informed Consent Form in Malayalam*	18. *Covering letter from Dr. Mathew Abraham dated 26.06.2020*

1

Revised documents

1. *Covering letter addressed to Chairman, IEC, SCTIMST dated 07/08/2020* 2. *TAC-Clinical Studies with comments included* 3. *Revised IEC Application form (not complete)* 4. *Revised Proposal* 5. *Revised proforma in English* 6. *Pain VAS score scale in English* 7. *Neck Disability index in English* 8. *Japanese Orthopedic Association Disfunctionality Score in English* *Chicago Chiari outcome scale in English* 9. *Scoliosis Patient Questionnaire Ver 30 in English* 10. *Pain VAS score scale in Malayalam* 11. *Neck Disability index in Malayalam* 12. *Japanese Orthopedic Association Disfunctionality Score in Malayalam*	13. *Chicago Chiari outcome scale in Malayalam* 14. *Scoliosis Patient Questionnaire Ver 30 in Malayalam* 15. *Patient information sheet in English* 16. *Patient information sheet in Malayalam* 17. *Consent form in English* 18. *Consent form in Malayalam* 19. *CV of PI Jayapalsinh Gohil with GMC Registration* 20. *CV of Co-PI Dr Ganesh Divakar with TCMC Registration* 21. *CV of Co-PI Dr Krishnakumar K with TCMC Registration* 22. *CV of Co-PI Dr Mathew Abraham with TCMC Registration* 23. *CV of Co-PI Dr Bejoy Thomas with TCMC Registration* 24. *CV of Co-PI Dr Harshavardhan Biradar with TNMC Registration*

The following members of the Ethics Committee were present at the meeting held onJuly 10-17, 2020 at the offices and residences of the members

SL. No.	Member Name	Highest Degree	Gender	Scientific /Non Scientific	Affiliation with Institution(s)
1.	Dr. R V G Menon	M Tech, PhD	Male	Lay Person (Chairman)	No
2.	Dr. Rema M. N	MD	Female	Basic Medical Scientist	No
3.	Dr. Kala Kesavan. P	MBBS, MD	Female	Basic Medical Scientist	No
4.	Smt. Sathi Nair	MA (English Literature)	Female	Lay Person	No
5.	Dr. Christina George	MD Psychiatry	Female	Clinician	No
6.	Dr. Harikrishnan S	MD, DM (Cardiology) DNB (Cardiology)	Male	Clinician	Yes
7.	Dr. Mala Ramanathan	PhD	Female	Social Scientist (Member Secretary)	Yes

IEC Decision

The IEC approved the conduct of the study in the present form.

Remarks:
The Institutional Ethics Committee expects to be informed about the progress of the study, any SAE occurring in the course of the study, any changes in the protocol and patient information/informed consent and asks to be provided a copy of the final report.

There was no member of the study team who participated in voting / decision making process. The ethics committee is organized and operated according to the requirements of Good Clinical Practice and the requirements of the Indian Council of Medical Research (ICMR).

Sincerely,

Mala Ramanathan
Member Secretary, IEC

2

Plagiarism Checker X Originality Report

Similarity Found: 8%

Date: Sunday, July 28, 2019
Statistics: 824 words Plagiarized / 10302 Total words
Remarks: Low Plagiarism Detected - Your Document needs Selective Improvement.

----------**Chiari malformation:**

"Chiari I malformation is defined as the descent of cerebellar tonsils below the foramen magnum evident on imaging. if descent is more than 5 mm for adult and more than 3 mm for a pediatric age group is the cut-off limit for definition. (1)"

. The Chiari I malformation (CIM) is the most common type, with a prevalence of 0.56% to 1.00% (1). Between 50% and 60% of CIM, patients are diagnosed with a concurrent condition, most commonly syringomyelia. Scoliosis is common not only in CIM but also in syrinx patients. Syringomyelia is regarded as a possible contributor to the pathogenesis of scoliosis in both CIM and idiopathic syringomyelia patients (2,3). Researchers have previously investigated the characteristics of syrinx and scoliosis in the CIM and idiopathic syrinx (IS) population separately (4) and there is no literature on differences between CIM and IS in terms of syrinx characteristics, scoliosis features, and clinical manifestations.

There are many studies on the outcome of Chiari malformation following foramen magnum decompression. However recently few authors have recommended C1-C2 fusion as an effective treatment of Chiari malformation and questioned the effectiveness of foramen magnum decompression. Our institute harbors the largest operated case number of Chiari malformation. We have done a retrospective analysis of clinical and radiological parameters changes following foramen magnum decompression.