Surgical Experience in Pediatric Patients with Chiari I malformations age ≤ 18 years

Submitted for MCh Neurosurgery

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CERTIFICATE

This is to certify that the thesis entitled “Surgical experience in pediatric patients with Chiari I malformations age ≤ 18 years” is a bonafide work of Dr. Vipin Kumar and was conducted in the Department of Neurosurgery, Sree Chitra Tirunal Institute for Medical Sciences & Technology, Thiruvananthapuram (SCTIMST), under my guidance and supervision.

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DECLARATION

This thesis titled “Surgical experience in pediatric patients with Chiari I malformations age ≤ 18 years” is a consolidated report based on a bonafide study of the period from January 1999 to June 2011, done by me under the Department of Neurosurgery, Sree Chitra Tirunal Institute for Medical Sciences & Technology, Thiruvananthapuram. This thesis is submitted to SCTIMST in partial fulfillment of rules and regulations of MCh Neurosurgery examination.

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INTRODUCTION

Chiari malformation Type I is characterized by caudal descent of cerebellar tonsils and may or may not be associated with the presence of a syrinx, a degree of medullary descent and buckling of the lower medulla may also be present. Herniation of the tonsils more than 5 mm below the foramen magnum on MR imaging is considered diagnostic in adults (1). Mikulis DJ. Et al - criteria for ectopia of the cerebellar tonsils: 1st decade of life, 6 mm; 2nd and 3rd decades, 5 mm; 4th to 8th decades, 4 mm; and 9th decade, 3 mm (40). It is more common in women, with a female-to-male ratio variably reported as 3:1 to 1:1 (28, 39). In pediatric chiari I malformation female-to-male ratio variably reported as 1.5:1 to 1:1.5 (20, 27, 59). If only patients with Chiari I malformation associated with syringomyelia are included, the female-to-male ratio ranges from 1:1 to 1.3 (29). The reported prevalence of tonsillar herniation extending more than 5 mm below the foramen magnum on MR imaging of the head is 0.78% (36). Some patients with this malformation are completely asymptomatic, and the diagnosis is established incidentally when MRI is performed for some other reason. Clinical manifestations – Headache common symptom of pediatric Chiari I malformation, is found in 40 – 60 % of patients (15, 27, 59, 61).
It is described as a heavy, crushing, or pressure-like sensation at the back of the head that radiates to the vertex, behind the eyes and to the neck and shoulders which accentuated by physical exertion and by Valsalva maneuvers (e.g., coughing, sneezing, or vomiting). Most patients also experience ocular disturbances, such as blurred vision, photophobia, diplopia, and visual field deficits. However, the neuro-ophthalmological examination in these patients usually is normal. 10-21% patients report a lower cranial nerve (15, 27, 59.). The most common symptoms in this group are dysphagia, sleep apnea, dysarthria, hoarseness. Syrinx reported in pediatric chiari I malformation ranges from 51-72% (27, 59, 61, 43, 44, 47, 48). The patients with syringomyelia more frequently suffered motor deficits than those without syringomyelia (ratio 7:1). Sensory symptoms were also more frequent in this group (ratio 4:1) (27). Associated diagnoses with pediatric chiari I malformation reported are hydrocephalus - 11%, type I neurofibromatosis - 5.5%, idiopathic growth hormone deficiency - 5.5%, Klippel–Feil anomaly - 5%, basilar invagination - 4% and Sprengel deformity - 2.7% (59, 61). The diagnosis of Chiari I malformation is suspected when analyzing the clinical course and physical examination, but it must be confirmed with imaging studies. MRI is considered the
most important study for establishing the diagnosis and planning the surgical treatment. A T1-weighted sagittal view of the CVJ usually shows both the tonsillar herniation and syringomyelia, but in patients with small spinal cord cavities, T2-weighted imaging also can be very helpful. MRI also is useful for identifying other related anomalies, such as tumors and CVJ problems. Dynamic MRI helps demonstrate abnormalities in CSF flow at the foramen magnum and the benefits of decompressive surgery in patients with the Chiari I malformation (57). Ultrasonography is another useful study in Chiari I malformation with mild Tonsillar herniation (rostral to C1), but it is used only during surgery to identify when the CSF circulation has been re-established during the procedure (34). The management strategy follows a “top down” rule (15, 42, 53, 58). This implies that one proceeds from the cranial to caudal direction. If there is hydrocephalus it is dealt with first by ventriculoperitoneal shunt or other appropriate shunting technique. If the shunting does not ameliorate the symptoms then foramen magnum decompression with a lax expansive duroplasty is done to deal with the impaction of tonsils into the upper cervical canal. The expansile duroplasty allows more room around the foramen magnum and opens the subarachnoid pathways. If with this manoeuvre the syrinx cavity
resolves or flow disappears then there is no need for further intervention. If the syrinx persists then one would consider direct shunting of the syrinx.

With the natural history known incompletely, poorest prognosis is seen in patients with central cord signs; the best prognosis is found in patients with paroxysmal intracranial hypertension (62). Saez et al (63) classify patients into preoperative prognostic categories revealed that more than 80% of patients with paroxysmal intracranial hypertension or cerebellar dysfunction achieved a favorable outcome, that 65% of patients with foramen magnum compression improved, and that only 33% of patients with central cord disturbance improved.
REVIEW OF LITERATURE

HISTORY:

Chiari malformations encompass a spectrum of congenital hindbrain herniation syndromes. First described by the Austrian pathologist Hans Chiari in 1891 (11). His work on type I Chiari malformation (CIM) was published in Deutsche Medizinische Wochenschrift. He described a 17-year-old girl who suffered from hydrocephalus but did not have symptoms attributable to the cerebellum or medulla. The patient died of typhoid fever. At autopsy, Chiari discovered an “elongation of the tonsils and medial parts of the inferior lobes of the cerebellum into cone-shaped projections, which accompany the medulla oblongata into the spinal canal.” Five years later, Chiari described 14 similar cases, noting that the grade of hydrocephalus was not related to the severity of the craniospinal changes. He theorized that an additional mechanism might play a role in this condition, such as insufficient bone growth and insufficient enlargement of skull parts, resulting in increased intracranial pressure. Briefly a few of the relevant historical vignettes in the description of this disease has been mentioned below:

1. 1901 First description of CIM-related neurological symptoms, by Home´n (22)
2. 1935 Russell and Donald (64) suggested that hydrocephalus could be secondary to the cranio-cervical deformity and could be treated by decompressing the foramen magnum. They also introduced the notion of Chiari malformation in the English-language literature;

3. 1938 McConnell and Parker (35) described the first adult patients with CIM (and hydrocephalus); they also used the term “tonsils” to indicate the prolapsed portion of the cerebellum. The same year, Aring (5) reported on the first case of CIM without hydrocephalus;

4. 1941 First radiological diagnosis of CIM, realized by Adams, Schatzki and Scoville (2). The authors reported on a patient with a “block” at the level of C3 on preoperative myelography. The authors also classified the symptoms of CIM into 5 groups:

(a) Raised intracranial pressure,
(b) Cranial nerve palsy,
(c) Brainstem compression,
(d) Spinal cord compression,
(e) Cerebellar signs

Regarding history of management of chiari I malformation:
Gardner (19) did decompression of the foramen magnum and atlas with opening of the fourth ventricle and plugging of the obex.
Williams (67) modified this technique by suturing muscle to the obex. This was based on the assumption that syringomyelic cavity in most patients communicated with the fourth ventricle through a patent obex. These obex plugging procedures were associated with a high risk of complications including bradycardia and hypotension besides a high failure rate. Hankinson (21) and later Peerless and Durward (52) advocated foramen magnum decompression with a fourth ventricle to cisterna magna drain. Bertrand (8) described subpial excision of the tonsils. Elimination of the craniospinal pressure dissociation forms the basis of most of the current surgical approaches. MR imaging studies have confirmed the validity of posterior fossa decompression as the initial operative approach to Chiari malformation. Rhoton (57) propagated foramen magnum decompression, establishing outlet from the fourth ventricle and upper cervical laminectomy with myelotomy. Logue and Edwards (33) performed foramen magnum decompression, left the dura open but kept the arachnoid intact. Isu, (23) removed the outer layer of dura along with foramen magnum decompression. Syrinx to cisternomagna shunt was advocated by Milhorat (40). The widely used procedure of foramen magnum decompression with a lax duroplasty has become the procedure of choice ever since Oldfield (50)
popularised his theory of formation of syrinx in cases associated with Chiari malformation.

**EMBRYOLOGY:**

All the structures of the brain develop from the neural tube. Fusion of the neural tube in the cranial region and closure of the rostral neuropore forms the three primary brain vesicles: forebrain (prosencephalon), midbrain (mesencephalon), and the hindbrain (rhombencephalon), by the 4th gestational week (49). Rhombencephalon further divides into two secondary vesicles by a marked constriction or “isthmus rhombencephali” into an upper vesicle called the metencephalon and the lower, the myelencephalon. The pons develops from a thickening in the floor and lateral walls of the metencephalon. The floor and lateral walls of the myelencephalon are thickened to form the medulla oblongata, which is continuous inferiorly with the spinal cord. The cerebellum is a derivative predominantly of the hindbrain but has a small contribution from the alar plates of the caudal third of the mesencephalon, which form the vermis. The alar plates of the metencephalon (cranial portion of the rhombencephalon) form the cerebellar hemispheres (68, 4). The cavity of the hindbrain becomes the future 4th ventricle.
The roof of the 4th ventricle is divided by the plica choroidalis into two areas: the anterior membranous area (AMA) superiorly and the posterior membranous area (PMA) inferiorly. Because of the neuroblastic proliferation, the alar laminae along the lateral margins of the AMA become thickened to form 2 lateral plates, or the rhombic lips. This process begins at approximately 4 to 6 weeks of gestation (7) and causes the rhombic lips to enlarge and then approach each other. Initially the rhombic lips (the developing cerebellum) project into the 4th ventricle, and as they enlarge and fuse in the median plane, they overgrow the rostral half of the 4th ventricle and overlap the pons and the medulla.

The structure of the cerebellum reflects its embryologic development (49). The archicerebellum (flocculonodular lobe), is the oldest part phylogenetically. It has connections with the vestibular apparatus. The paleocerebellum (vermis and anterior lobe) is of a more recent phylogenetic development. It is associated with density data from the limbs. The neocerebellum (posterior lobe), is the newest part phylogenetically. This portion of the cerebellum is associated with selective control of limb movements. Disorders of genes involved in early cerebellar patterning have been shown to produce a wide spectrum
of abnormal phenotypes, ranging from agenesis of the entire midbrain cerebellar region to minimal abnormalities of cerebellar foliation (68).

As the rhombic lips grow, the AMA regresses. The AMA is completely incorporated into the developing choroid plexus. Growth and backward extension of the cerebellum pushes the choroid plexus inferiorly, whereas the PMA greatly diminishes in the relative size compared with the overgrowing cerebellum. Subsequently there is development of a marked caudal protrusion of the 4th ventricle, causing the PMA to expand as the finger of a glove (13).
This transient protrusion has been labeled Blake’s pouch (51). The Blake’s pouch consists of ventricular ependyma surrounded by condensation of the mesenchymal tissues (13). The Blake’s pouch is initially a closed cavity that does not communicate with the surrounding subarachnoid space of the cisterna magna. The network between the vermis and the Blake’s pouch progressively becomes condensed, where as the other portions about the evagination become rarified and thus permeabilization of the Blake’s pouch occurs, which then forms the foramen of Magendie. The formation of the cisterna magna presumably occurs at approximately 6 weeks of gestation (14), which is in
communication with the 4th ventricle via the foramen of Magendie. The precise time of the opening of the foramen of Magendie is not established; however, persistence of the Blake’s pouch has been demonstrated into the 4th gestational month (56). The foramina of Luschka also probably appear late into the 4th month of gestation (4).

ANATOMY OF CVJ:

Structures involved in CVJ lesions include the lower cranial and upper spinal nerves, the caudal brainstem and rostral spinal cord, the vertebral artery and its branches, the veins and dural sinuses at the craniovertebral junction, and the ligaments and muscles uniting the atlas, axis, and occipital bone (3).

Osseous Relationship:

Occipital bone:

The occipital bone surrounds the foramen magnum. The foramina opening is oval shaped and is wider posteriorly than anteriorly. The wider posterior part transmits the medulla, and the narrower anterior part sits above the odontoid process. The occipital bone is divided into a squamosal part located above and behind the foramen magnum, a basal part situated in front of the foramen magnum, and paired condylar parts located lateral to the foramen magnum. The occipital condyles, which
articulate with the atlas, protrude from the external surface of this part. These condyles are located lateral to the anterior half of the foramen magnum. They are oval in shape, convex downward, face downward and laterally, and have their long axes directed forward and medially. A tubercle that gives attachment to the alar ligament of the odontoid process is situated on the medial side of each condyle.
The atlas:

The atlas, the first cervical vertebra, differs from the other cervical vertebrae by being ring shaped and by lacking a vertebral body and a spinous process. It consists of two thick lateral masses situated at the anterolateral parts of the ring. The lateral masses are connected in front by a short anterior arch and behind by a longer curved posterior arch. The position of the usual vertebral body is occupied by the odontoid process of the axis. Arch is convex backward and has a median posterior tubercle and a groove on the lateral part of its upper-outer surface in which the vertebral artery courses the arch. The first cervical spinal nerve also lies in groove, which is located between artery and the bone.

Figure. 4
The axis:

The axis, the second cervical vertebra, more closely resembles the typical vertebrae than the atlas, but is distinguished by the odontoid process (dens), which projects upward from the body. On the front of the dens is an articular facet that forms a joint with the facet on the back of the anterior arch of the atlas. The dens has a pointed apex that is joined by the apical ligament, has a flattened side where the alar ligaments are attached, and is grooved at the base of its posterior surface where the transverse ligament of the atlas passes. The dens and body are flanked by a pair of large oval facets that extend laterally from the body onto the adjoining parts of the pedicles and articulate with the inferior facets of the atlas.

Figure 5
Muscular relationships:
The craniovertebral junction is surrounded by the muscles attached to the occipital bone and upper cervical vertebrae. The trapezius covers the back of the head and neck. It extends from the medial half of the superior nuchal line, the external occipital protuberance, and the spinous processes of the cervical and thoracic vertebrae and converges on the shoulder to attach to the scapula and the lateral third of the clavicle. The sternocleidomastoid passes obliquely downward across the side of the neck from the lateral half of the superior nuchal line and mastoid process to the upper part of the sternum and the adjacent part of the clavicle. The splenius capitis, situated deep to and partially covered by the trapezius and sternocleidomastoid, extends from the bone below the lateral third of the superior nuchal line to the spinous processes of the lower cervical and upper thoracic vertebrae. The semispinalis capitis, which attaches above in the area between the superior and inferior nuchal lines beginning medially at the external occipital crest and extending laterally to the occipitomastoid junction, and the longissimus capitis muscle, which attaches above to the posterior margin of the mastoid process. The suboccipital muscles includes the superior oblique, which extends from the area lateral to the semispinalis capitis
between the superior and inferior nuchal lines to the transverse process of the atlas; the inferior oblique, which extends from the spinous process and lamina of the axis to the transverse process of the atlas; the rectus capitis posterior major, which extends from and below the lateral part of the inferior nuchal line to the spine of the axis; and the rectus capitis posterior minor, which is situated medial to and is partially covered by the rectus capitis posterior major, extends from the medial part and below the inferior nuchal line to the tubercle on the posterior arch of the atlas. The anterior vertebral muscles insert on the clival part of the occipital bone anterior to the foramen magnum. This group includes the longus colli, which attach to the anterior surface of the vertebral column between the atlas and the third thoracic vertebra; the longus capitis, which extends from the clivus in front of the foramen magnum to the transverse processes of the third through the sixth cervical vertebrae; the rectus capitis anterior, which is situated behind the upper part of the longus capitis and extends from the occipital bone in front of the occipital condyle to the anterior surface of the lateral mass and transverse process of the atlas; and the rectus capitis lateralis, which extends from the jugular process of the occipital bone to the transverse process of the atlas.
Neural relationships:

The neural structures situated in the region of the craniocervical junction are the caudal part of the brainstem, cerebellum and fourth ventricle, the rostral part of the spinal cord, and the lower cranial and upper cervical nerves.

Cervicomedullary Junction:

The spinal cord blends indistinguishably into the medulla at a level arbitrarily set to be at the upper limit of the dorsal and ventral rootlets forming the first cervical nerve. The fact that the junction of the spinal cord and medulla is situated at the rostral margin of the first cervical root means that the medulla, and not the spinal cord, occupies the foramen magnum. At the craniocervical junction, the dentate ligament is located between the vertebral artery and the ventral roots of C1 anteriorly and the branches of the posterior spinal artery and the spinal accessory nerve posteriorly. The most rostral attachment of the dentate ligament is located at the level of the foramen magnum, above where the vertebral artery pierces the dura. The ventral root is located anterior to the dentate ligament, and the dorsal root, which is infrequently present, passes posterior to the dentate ligament.
**Brainstem:**

The lower medulla blends indistinguishably into the upper spinal cord at the level of the C1 nerve roots. The anterior surface of the medulla is formed by the medullary pyramids, which face the clivus, the anterior edge of the foramen magnum, and the rostral part of the odontoid process. The lateral surface is formed predominantly by the inferior olives. The posterior surface of the medulla is divided into superior and inferior parts. The superior part is composed in the midline of the inferior half of the fourth ventricle, and laterally by the inferior cerebellar peduncles. The inferior part of the posterior surface is composed of the gracile fasciculus and tubercle medially, and the cuneate fasciculus and tubercle laterally.

**Cerebellum:**

The sub occipital cerebellar surface rests above the posterior and lateral edge of the foramen magnum. Only the lower part of the hemispheres formed by the tonsils and the biventral lobules, and the lower part of the vermis formed by the nodule, uvula, and pyramid, are related to the foramen magnum. The biventral lobule sits above the lateral part of the foramen magnum, and the tonsils rest above the level of the posterior edge. The cerebellar surface above the posterior part of the foramen
magnum has a deep vertical depression, the posterior cerebellar incisura, which contains the falx cerebelli and extends inferiorly toward the foramen magnum. The tonsils, which sit above the posterior edge of the foramen magnum, are commonly involved in herniation through the foramen magnum. Each tonsil is an ovoid structure that is attached along its superolateral border to the remainder of the cerebellum. The cerebellomedullary fissure extends superiorly between the cerebellum and the medulla and is situated rostral to the posterior margin of the foramen magnum.

**Cranial nerves:**

The accessory nerve is the only cranial nerve that passes through the foramen magnum. It has a cranial part composed of the rootlets that arise from the medulla and join the vagus nerve, and a spinal portion formed by the union of a series of rootlets that arise from the lower medulla and upper spinal cord. In the posterior fossa, the accessory nerve is composed of one main trunk from the spinal cord and three to six small rootlets that emerge from the medulla. The lower four cranial nerves are sufficiently close to the foramen magnum that they may be involved by lesions arising there.
**Cervical nerve roots:**

Each dorsal and ventral root is composed of a series of six to eight rootlets that fan out to enter the posterolateral and anterolateral surfaces of the spinal cord, respectively. The dorsal and ventral roots cross the subarachnoid space and transverse the dura mater separately, and then unite close to the intervertebral foramen to form the spinal nerves. The rootlets in the region of the foramen magnum pass almost directly lateral to reach their dural foramina. The neurons of the dorsal roots collect to form ganglia located just proximal to the union of the dorsal and ventral root in the intervertebral foramina, however the first cervical dorsal root and associated ganglion may be absent. The C1, C2, and C3 nerves, distal to the ganglion, divide into dorsal and ventral rami. The dorsal rami divide into medial and lateral branches that supply the skin and muscles of the posterior region of the neck. The C1 nerve, termed the sub occipital nerve, leaves the vertebral canal between the occipital bone and atlas and has a dorsal ramus that is larger than the ventral ramus. The dorsal ramus courses between the posterior arch of the atlas and the vertebral artery to reach the sub occipital triangle, where it sends branches to the rectus capitis posterior major and minor, superior and inferior oblique, and the semispinalis capitis, and occasionally has a
cutaneous branch that accompanies the occipital artery to the scalp. The C1 ventral ramus courses between the posterior arch of the atlas and the vertebral artery and passes forward, lateral to the lateral mass of the atlas and medial to the vertebral artery, and supplies the rectus capitis lateralis. The C2 nerve passing below and supplying the inferior oblique muscle, the dorsal ramus divides into a large medial and a small lateral branch. It is the medial branch that is most intimately related to this suboccipital operative field and that forms the greater occipital nerve. It ascends obliquely between the inferior oblique and the semispinalis capitis, pierces the latter and the trapezius muscle near their attachments to the occipital bone, and is joined by a filament from the medial branch of C3. It supplies the semispinalis capitis muscle, ascends with the occipital artery, and supplies the scalp as far forward as the vertex, and occasionally the back of the ear. The lateral branch sends filaments that innervate the splenius, longissimus, and semispinalis capitis, and is often joined by the corresponding branch from the C3 nerve. The C2 ventral ramus courses between the vertebral arches and transverse processes of the atlas and axis and behind the vertebral artery to leave this operative field. Two branches of the C2 and C3 ventral rami, the lesser occipital and greater auricular nerves, curve around the posterior
border and ascend on the sternocleidomastoid muscle to supply the skin behind the ear.

**Arterial relationships:**
The major arteries related to the foramen magnum are the vertebral and posteroinferior cerebellar arteries (PICA), and the meningeal branches of the vertebral, and external and internal carotid arteries.

**Vertebral artery:**
The paired vertebral arteries arise from the subclavian arteries, ascend through the transverse processes of the upper six cervical vertebrae, pass behind the lateral masses of the axis, enter the dura mater behind the occipital condyles, ascend through the foramen magnum to the front of the medulla, and join to form the basilar artery at the pontomedullary junction. Each artery is divided into intradural and extradural parts. The extradural part is divided into three segments. The first segment extends from the origin at the subclavian artery to the entrance into the lowest transverse foramen, usually at the C6 level. The second segment ascends through the transverse foramina of the upper six cervical vertebrae in front of the cervical nerve roots. This segment deviates laterally just above the axis to reach the laterally placed transverse foramen of the atlas. The third segment, the one most intimately related to the foramen magnum, extends from the foramen in the transverse process of the atlas
to the site of passage through the dura mater. The third segment passes medially behind the lateral mass of the atlas and atlanto-occipital joint and is pressed into the groove on the upper surface of the lateral part of the posterior arch of the atlas. It enters the vertebral canal by passing anterior to the lateral border of the atlantooccipital membrane. It is surrounded by a venous plexus composed of anastomoses between the deep cervical and epidural veins. The C1 nerve root passes through the dura mater on the lower surface of the vertebral artery between the artery and the groove on the posterior arch of the atlas with the vertebral artery. The terminal extradural segment of the vertebral artery gives rise to the posterior meningeal and posterior spinal arteries, branches to the deep cervical musculature, and infrequently the PICA. The intradural segment begins at the dural foramina just inferior to the lateral edge of the foramen magnum. Once inside the dura mater, the artery ascends from the lower lateral to the upper anterior surface of the medulla. The intradural part of the artery is divided into lateral and anterior medullary segments. The lateral medullary segment begins at the dural foramen and passes anterior and superior along the lateral medullary surface to terminate at the preolivary sulcus. The anterior medullary segment begins at the preolivary sulcus, courses in front of, or between, the
hypoglossal rootlets, and crosses the pyramid to join with the other vertebral artery at or near the pontomedullary sulcus to form the basilar artery. The branches arising from the vertebral artery in the region of the foramen magnum are the posterior spinal, anterior spinal, PICA, and anterior and posterior meningeal arteries.

**Posteroinferior cerebellar artery:**

The PICA is the largest branch of the vertebral artery. It usually originates with the dura mater, but it may infrequently originate from the terminal extradural part of the vertebral artery. It may arise at, above, or below the level of the foramen magnum. The tonsillomedullary PICA segment, which forms the caudal loop related to the lower part of the tonsil, is most intimately related to the foramen magnum.

**Anterior spinal artery:**

The anterior spinal artery is formed by the union of the paired anterior ventral spinal arteries, which originate from the anterior medullary segment of the vertebral arteries near the origin of the basilar artery. The anterior spinal artery descends through the foramen magnum on the anterior surface of the medulla and the spinal cord in or near the anteromedian fissure. On the medulla, it supplies the pyramids and their decussation, the medial lemniscus, the interolivary bundles, the
hypoglossal nuclei and nerves, and the posterior longitudinal fasciculus. It anastomoses with the anterior branches of the radicular arteries entering the cervical foramina. There are few anastomoses with the anterior radicular branches if the descending channel is large, but it has frequent connections with the anterior radicular arteries if it is small.

Meningeal arteries:
The dura mater around the foramen magnum is supplied by the anterior and posterior meningeal branches of the vertebral artery, and the meningeal branches of the ascending pharyngeal and occipital arteries. The anterior meningeal branch of the vertebral artery arises from the medial surfaces of the extradural part of the vertebral artery immediately above the transverse foramen of the third cervical vertebra. The artery enters the spinal canal through the intervertebral foramen between the second and third cervical vertebrae, and ascends between the posterior longitudinal ligament and the dura mater. At the level of the apex of the dens, each artery courses medially to join its mate from the opposite side and forms an arch over the apex of the dens. Its branches supply the dura mater in the region of the clivus and the anterior part of the foramen magnum and upper spinal canal, and they anastomose with the branches of the ascending pharyngeal and dorsal meningeal arteries that
supply the dura mater covering the anterior and anterolateral part of the posterior fossa. The anterior meningeal artery also gives rise to muscular and osseous branches that supply the body and odontoid process of the axis and the articulate plate of the atlanto-occipital and atlantoaxial joints. The posterior meningeal artery arises from the posterosuperior surface of the vertebral artery as it courses around the lateral mass of the atlas, above the posterior arch or just before penetrating the dura. After passing through the foramen magnum, it ascends near the falx cerebelli and divides near the torcula into several branches that terminate in the posterior part of the tentorium and cerebral falx. It supplies the dura mater lining the posterolateral and posterior part of the posterior cranial fossa, and anastomoses with the meningeal branches of the ascending pharyngeal and occipital arteries. The ascending pharyngeal branch of the external carotid artery usually sends two branches to the dura above the foramen magnum. One branch passes through the hypoglossal canal and the other enters through the jugular foramen. The branch passing through the hypoglossal canal divides into an ascending branch that passes upward in the dura covering the clivus and anastomoses with the branches of the dorsal meningeal artery, and a descending branch that courses inferomedially toward the anterior edge of the foramen magnum.
and anastomoses with branches of the arcade above the odontoid process formed by the anterior meningeal arteries. The branches that enter through the jugular foramen divide into branches that course posteriorly and posterosuperiorly to anastomose with the meningeal branches of the occipital and posterior meningeal arteries, and supply the dura mater in the posterior and posterolateral parts of the posterior cranial fossa. The meningeal branch of the occipital artery is inconstant and, if present, it penetrates the cranium through the mastoid emissary foramen. It divides into one branch that courses posterosuperiorly to join the branches of the posterior meningeal artery that supplies the dura mater in the posterior part of the posterior fossa, and another branch that courses anterolaterally and joins the meningeal branches of the ascending pharyngeal artery.

**Venous relationships**

The venous structures in the region of craniocervical junction are divided into three groups: one composed of the extradural veins, another formed by the intradural (neural) veins, and a third constituted by the dural venous sinuses. The three groups anastomose through bridging and emissary veins. Extradural groups - Venous flow in this area empties into two systems: one drained by the internal jugular vein and
another draining into the vertebral venous plexus. The internal jugular vein and its tributaries form the most important drainage system in the craniocervical area. The internal jugular vein originates at the jugular foramen by the confluence of the sigmoid and inferior petrosal sinuses. The venous plexus surrounding the vertebral artery in the sub occipital triangle is formed by numerous small channels that empty into the internal vertebral plexuses (between the dura and the vertebrae), which issue from the vertebral canal above the posterior arch of the atlas. The posterior condylar emissary vein, which passes through the posterior condylar canal, forms a communication between the vertebral venous plexus and the sigmoid sinus. The venous plexus of the hypoglossal canal passes along the hypoglossal canal to connect the basilar venous plexus with the marginal sinus, which encircles the foramen magnum. The venous channels in the dura mater surrounding the foramen magnum are the marginal, occipital, sigmoid, inferior petrosal, and basilar venous plexus. The marginal sinus is located between the layers of the dura in the rim of the foramen magnum. It communicates anteriorly, through a series of small sinuses, with the basilar sinus on the clivus, and posteriorly with the occipital sinus. It is usually connected to the sigmoid sinus or jugular bulb, by a sinus that passes across the
intracranial surface of, and communicates with, the veins in the hypoglossal canal. These anastomoses provide an alternative route for venous drainage in the case of obstruction of the internal jugular vein.

The occipital sinus courses in the cerebellar falx. Its lower end divides into paired limbs each of which courses anteriorly around the foramen magnum to join the sigmoid sinus or the jugular bulb and its upper end joins the torcula. The basilar venous plexus is located between the layers of the dura mater on the upper clivus. It is formed by interconnecting venous channels that anastomose with the inferior petrosal sinuses laterally, the cavernous sinuses superiorly, and the marginal sinus and epidural venous plexus inferiorly. The intradural veins in the region of the foramen magnum drain the lower part of the cerebellum and brainstem, the upper part of the spinal cord, and the cerebellomedullary fissure. The veins of the medulla and spinal cord form longitudinal plexiform channels that anastomose at the foramen magnum. The median anterior spinal vein that courses in the anteromedian spinal fissure deep to the anterior spinal artery is continuous with the median anterior medullary vein that courses on the anteromedian sulcus of the medulla. The lateral anterior spinal vein courses longitudinally along the origin of the ventral roots and superiorly joins the lateral anterior
medullary vein that courses longitudinally in the anterolateral medullary (preolivary) sulcus along the line of origin of the hypoglossal rootlets. The lateral posterior spinal vein, which courses along the line of origin of the dorsal roots in the posterior lateral spinal sulcus, is continuous above with the lateral medullary vein that courses along the retro-olivary sulcus, dorsal to the olive. The median posterior spinal vein, which courses along the posteromedian spinal sulcus, is continuous above with the main vein on the posterior surface of the medulla, the median posterior medullary vein that courses along the posteromedian medullary sulcus. The transverse medullary and transverse spinal veins cross the medulla and spinal cord at various levels, interconnecting the major longitudinal channels. Bridging veins may connect the neural veins with the dural sinus in the region of the foramen magnum
DEFINITIONS (62):

**Chiari Type 0:** The Chiari 0 malformation is defined as syringomyelia without tonsillar herniation that responds to posterior fossa decompression.

**Chiari Type I:** Tonsillar herniation >5 mm inferior to the plane of the foramen magnum (basion – opisthion line).

No associated brainstem herniation or supratentorial anomalies. Hydrocephalus is uncommon. Syringomyelia is common.

**Chiari Type 1.5 (60):** Chiari I malformation is seen in combination with brainstem herniation (an inferiorly displaced obex beneath the foramen magnum (basion–opisthion line))

**Chiari Type II:** Herniation of the cerebellar vermis, brainstem, and fourth ventricle through the foramen magnum. Associated with myelomeningocele and multiple brain anomalies. Hydrocephalus and syringomyelia very common

**Chiari Type III:** High cervical or occipital encephalocele containing herniated cerebellar and brainstem tissue

**Chiari Type IV:** Hypoplasia or aplasia of the cerebellum and tentorium cerebelli
Tonsillar herniation:

In 1963, Baker (6) proposed that the normal position of cerebellar tonsils is above a line joining the tip of the clivus and the posterior rim of the foramen magnum (basion–opisthion line). About 10 years later, Bloch et al. (9) defined the myelographic position of the tonsils in normal and in CIM subjects. In asymptomatic individuals, this position ranged from 7 mm above to 8 mm below the foramen magnum, while it ranged from 3 mm above to 25 mm below the foramen in symptomatic subjects. Therefore, the authors concluded that “tonsillar herniation can be seen as an incidental finding”. The advent of MRI radically changed the way to obtain the diagnosis of CIM. In 1985, Aboulezz et al. (1) investigated 82 normal subjects and 11 CIM individuals by means of MRI: the position of the tonsils ranged from 20 mm above to 2.8 mm below the foramen in normal individuals, and from 5.2 to 17.7 mm below the foramen in affected patients, whose tonsils also appeared as pointed. The authors considered tonsils up to 3 mm under the foramen as normal, and those more than 5 mm below as indicative of CIM. Similarly, Barkovich et al. (7) noticed that the position of the tonsils among 200 asymptomatic controls varied from 8 mm above to 5 mm below the foramen magnum, while the tonsillar herniation among 25
CIM patients was 3–29 mm below the foramen, so that they indicated 5 mm as the normal limit under the foramen. According to Elster and Chen (17), the 5 mm cut-off is valid in case of ectopia of one tonsil, while a 3–5 mm cut-off is more appropriate for the herniation of both tonsils (borderline CIM).

**PATHOPHYSIOLOGY:**

**TONSILLAR HERNIATION:** Tonsillar herniation may be congenital or acquired. Chiari's belief that hydrocephalus causes Tonsillar herniation has been abandoned because it is present in only a minority of cases (62). The underdevelopment of occipital somites within the paraxial mesoderm creates a small posterior fossa and CIM. This contention is supported by the association of CIM with other spine, skull, somatic, and craniofacial abnormalities, which are the result of mesodermal maldevelopment (41). The association of craniosynostosis and CIM is known and appears strongest in cases of syndromic, multisuture, and lambdoid synostosis (12). Cephalocranial disproportion in multisuture synostosis can elevate intracranial pressure and promote herniation of posterior fossa elements. Medical conditions may promote formation of an abnormally small posterior fossa. Familial vitamin D-resistant rickets causes bony overgrowth of the posterior fossa, thus
reducing its volume (32, 62). Development of a craniospinal pressure
gradient across the foramen magnum may cause or hasten the
development of Tonsillar herniation. The gradient results from impaired
CSF flow across the foramen magnum. Negative CSF pressure in the
spinal compartment relative to the intracranial compartment creates a
"sump effect" that forces the tonsils down through the foramen
magnum. Once CSF flow is blocked at the foramen magnum, low
intraspinal pressures can be accentuated and perpetuated by continuous
absorption of CSF through spinal pathways, further worsening the
clinical situation. Lumboperitoneal shunting, repetitive lumbar
punctures, lumbar drainage, and chronic spinal CSF leaks of an
iatrogenic nature are causes of an acquired tonsillar herniation (62).

SYRINGOMYELIA:

In the 1960s, Gardner (19) presented the hydrodynamic theory.
Gardner's theory stated that in normal embryology, CSF pulsations from
the choroid plexus play a significant role in the expansion of the neural
tube. These pulsations help with the development of the arachnoid
pathways as well as with modeling of the expanding brain. The balance
between the pulsatile flow in the supratentorial and fourth ventricular
choroid plexus directed brain growth differentially: if the fourth
ventricular pulsations were overactive, the tentorium would be pushed upward, and a Dandy-Walker malformation could develop. Conversely, if the supratentorial pulsations were overactive, tentorial migration becomes such that the posterior fossa is small, allowing the development of a Chiari malformation; in addition, the CSF outlets of the fourth ventricle would remain closed, directing the CSF into the patent opening at the obex, thus causing syringomyelia.

Williams (67) expanded on Gardner's theory by suggesting that Valsalva maneuvers resulted in epidural venous congestion and intracranial as well as intraspinal pressure elevation, causing fluid to flow both cranially and caudally. Although flow into the cranial compartment meets no resistance, caudal flow is delayed by hindbrain adhesions and outlet obstruction, thus creating a pressure differential between the cranial and spinal compartments. This pressure differential may last a few seconds and cause worsening hindbrain impaction and syringomyelia. Repeat measurements were made after surgical decompression, showing equilibration of the pressures in the two compartments, which, in turn, correlated with clinical improvement. However, spinal cord cavitation is often acquired (as in posttraumatic syringomyelia), and a connection between the cyst and the fourth
ventricle is not always present, which raises doubts about the adequacy of this theory.

Oldfield (50) and associates investigated the anatomy and dynamics of movement of the cerebellar tonsils and CSF during the respiratory and cardiac cycles to explore the mechanism of syringomyelia progression in patients with CIM. During systole, there is normally movement of CSF in a caudal direction across the foramen magnum to counter the increased intracranial volume of blood and maintain physiologic intracranial pressure. This flow reverses in diastole. Dynamic movement of subarachnoid fluid is mirrored by caudal and cranial pulsations of fluid within the central canal during systole and diastole, respectively. In patients with CIM, the cerebellar tonsils are forced down and obstruct CSF flow across the foramen magnum during systole. This piston-like movement of the cerebellar tonsils imparts a systolic pressure wave in the spinal CSF that acts on the surface of the spinal cord, forcing fluid into the cord through perivascular and interstitial spaces. Oldfield and collaborators demonstrated the dynamic CSF flow into the syrinx in patients with CIM preoperatively by dynamic MRI and intraoperative by ultrasound and further demonstrated the resolution of pathologic flow following bony and dural decompression. Postoperatively, adequate
decompression of the foramen magnum allows resolution of the syringomyelia.

**CLINICAL FINDINGS:**

Symptoms of Chiari malformations are related to age: in infancy, signs of brainstem compression predominate with apnea spells, cyanosis attacks, and swallowing problems, whereas in later childhood scoliosis becomes the most common presenting sign. The typical clinical features of occipital headache, gait ataxia, sensory disturbances, and motor weakness are uncommon in children and observed predominantly in adults. The age-related clinical course may be explained by the postnatal growth of the cerebellum. At birth, most parts of the brain have reached about a third of their adult volume. The cerebellum is the smallest part of the central nervous system with just 15% of its adult volume at this time, presumably to protect the brainstem during delivery. The adult volume of the cerebellum is reached in the second year of life, indicating that the cerebellar volume increases by a factor of 7 in that period. This may explain the dramatic presentations with respiratory problems in this age group, something almost unknown to adult patients. Once the cerebellum is outgrown, the clinical course tends to be characterized by slow progression (25).
Several authors have broadly divided signs and symptoms into frequently encountered clinical syndromes:

1) **Brain stem and bulbar palsy syndrome**: a complex yet frequent presentation consisting of signs and symptoms attributable to lesions at the craniovertebral junctions, including variable involvement of the cranial nerves, lower brain stem, and/or cervical cord. Symptoms include headache, vertigo, ataxia, nystagmus, dysphagia, motor weakness, and variable long tract signs. Saez et al. (63) 38.3% of cases presented with findings of foramen magnum compression. K. S. Paul et al. (28) 22% cases presented with findings of foramen magnum compression.

2) **Paroxysmal intracranial hypertension**: consisting of symptoms that include exertional headache and nausea, vomiting, and dizziness associated with a headache episode. Neurologic examination frequently reveals normal or very mild signs. Saez et al. (63) 21.7% of cases presented with findings of paroxysmal intracranial hypertension.

3) **Central cord syndrome**: consisting of signs and symptoms attributable to lesions of the cervical cord, including pain (frequently “burning”), dissociated and posterior column sensory loss, segmental weakness or wasting, and long tract signs. The clinical impression of
these cases is almost invariably that of syringomyelia or intramedullary tumor. Saez et al, (63) 20% of cases presented with findings of central cord syndrome. K. S. Paul et al, (28) 65% of cases presented with findings of central cord syndrome.

4) **Cerebellar syndrome**: consisting of signs and symptoms attributable to lesions of the cerebellum, including uni or bilateral ataxia of the limbs or trunk, nystagmus, and dysarthria. Saez et al, (63) 10% of cases presented with cerebellar dysfunction. K. S. Paul et al, (28) 11% of cases presented with cerebellar dysfunction.

5) **Pyramidal syndrome**: consisting of signs and symptoms of stiffness and/or spasticity, and hyperreflexia. Saez et al, (63) 6.7% of cases presented with spasticity.

6) **Bulbar Palsy**: Presentation with isolated impairment of lower cranial nerve function. Saez et al, (63) 3.3% of cases presented with bulbar palsy.

Spontaneous Resolution of Chiari I Malformation and Syringomyelia is also reported (26). The common associated diagnosis includes scoliosis (18%), hydrocephalus (9.6%), type I neurofibromatosis (5%), idiopathic growth hormone deficiency (4.2%), basilar invagination (3%) and Sprengel deformity (0.8%) (59).
Nair et al, (46) proposed a grading system to evaluate the correlation between pre and post operative clinical and radiological parameters and to prognosticate its outcome.

**Clinical Scoring System(46)**

<table>
<thead>
<tr>
<th>Clinical feature</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Pain</strong></td>
<td></td>
</tr>
<tr>
<td>Absent</td>
<td>0</td>
</tr>
<tr>
<td>Mild-moderate</td>
<td>1</td>
</tr>
<tr>
<td>Incapacitating</td>
<td>2</td>
</tr>
<tr>
<td><strong>Sensory</strong></td>
<td></td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>0</td>
</tr>
<tr>
<td>Symptoms + No deficit</td>
<td>1</td>
</tr>
<tr>
<td>±Symptoms + &lt; 50 % deficit</td>
<td>2</td>
</tr>
<tr>
<td>±Symptoms + &gt; 50 % deficit</td>
<td>3</td>
</tr>
<tr>
<td><strong>Cranial Nerve</strong></td>
<td></td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>0</td>
</tr>
<tr>
<td>Symptoms + No deficit</td>
<td>1</td>
</tr>
<tr>
<td>±Symptoms + Unilateral deficit</td>
<td>2</td>
</tr>
<tr>
<td>±Symptoms + Bilateral deficit</td>
<td>3</td>
</tr>
<tr>
<td><strong>Cerebellum</strong></td>
<td></td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>0</td>
</tr>
<tr>
<td>Symptoms + No deficit</td>
<td>1</td>
</tr>
<tr>
<td>±Symptoms + Unilateral deficit</td>
<td>2</td>
</tr>
<tr>
<td>±Symptoms + Bilateral deficit</td>
<td>3</td>
</tr>
<tr>
<td><strong>Motor</strong></td>
<td></td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>0</td>
</tr>
<tr>
<td>Proximal/Distal only</td>
<td></td>
</tr>
<tr>
<td>Grade IV</td>
<td>1</td>
</tr>
<tr>
<td>Grade III</td>
<td>2</td>
</tr>
<tr>
<td>Grade II/I</td>
<td>3</td>
</tr>
<tr>
<td><strong>Proximal/Distal both</strong></td>
<td></td>
</tr>
<tr>
<td>Grade IV</td>
<td>1</td>
</tr>
<tr>
<td>Grade III</td>
<td>2</td>
</tr>
<tr>
<td>Grade II/I</td>
<td>3</td>
</tr>
</tbody>
</table>
Clinical Grade (63)

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Asymptomatic</td>
<td>Score 0</td>
</tr>
<tr>
<td>I</td>
<td>Mild</td>
<td>Score 1-5</td>
</tr>
<tr>
<td>II</td>
<td>Moderate</td>
<td>Score 6-10</td>
</tr>
<tr>
<td>III</td>
<td>Severe</td>
<td>Score 11-15</td>
</tr>
</tbody>
</table>

Klekamp J et al (25, 30) documented for individual symptoms according to a scoring system.

<table>
<thead>
<tr>
<th>Score</th>
<th>Pain</th>
<th>Sensory Dist., Dysesthesias</th>
<th>Motor Weakness</th>
<th>Gait Ataxia</th>
<th>Sphincter Function</th>
<th>Swallowing Function</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>None</td>
<td>Normal</td>
<td>Full power</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>4</td>
<td>Slight, no medication</td>
<td>Present, not significant</td>
<td>Movement against resistance</td>
<td>Unsteady, no aid</td>
<td>Slight dist., no catheter</td>
<td>Discomfort</td>
</tr>
<tr>
<td>3</td>
<td>Good control with medication</td>
<td>Significant, function not restricted</td>
<td>Movement against gravity</td>
<td>Mobile with aid</td>
<td>Residual, no catheter</td>
<td>Nerve deficit compensated</td>
</tr>
<tr>
<td>2</td>
<td>Insufficient control with medication</td>
<td>Some restriction of function</td>
<td>Movement without gravity</td>
<td>Few steps with aid</td>
<td>Rarely incontinent</td>
<td>Aspiration, coughing preserved</td>
</tr>
<tr>
<td>1</td>
<td>Severe despite medication</td>
<td>Severe restriction of function</td>
<td>Contraction without movement</td>
<td>Standing with aid</td>
<td>Often catheter</td>
<td>Aspiration, coughing insufficient</td>
</tr>
<tr>
<td>0</td>
<td>Incapacitating</td>
<td>Incapacitated function</td>
<td>Plegia</td>
<td>Plegia</td>
<td>Permanent catheter</td>
<td>No function</td>
</tr>
</tbody>
</table>
IMAGING:

MRI is the most important study for establishing the diagnosis and planning the surgical treatment. A T1-weighted sagittal view of the CVJ usually shows both the tonsillar herniation and syringomyelia, but in patients with small spinal cord cavities, T2-weighted imaging also can be very helpful. Axial images shows location of syrinx (central/eccentric) and maximum diameter of syrinx. R. S. Tubbs et al. (59) classified syrinx as small (maximum diameter less than one-third the diameter of the cord), medium (maximum diameter equal to half the diameter of the cord) and large (maximum diameter greater than half the diameter of the cord). CSF flow studies demonstrate impaired CSF flow across the foramen magnum. Rafeeqeque A et al. (55) demonstrated impaired systolic and unaffected diastolic CSF flow pulsations immediately below the foramen magnum in patients with CM 1. The systolic flow pulsations increase after surgery, and a correlation was established between postoperative improvement and changes in the CSF flow waveforms. Ultrasonography is another useful study in Chiari I malformation with mild Tonsillar herniation (rostral to C1), but it is used only during surgery to identify when the CSF circulation has been re-established during the procedure (34). Matthew J. Mcgirt et al,
demonstrated moderate-to-severe tonsillar CM-I, intraoperative ultrasonography demonstrating decompression of the subarachnoid spaces ventral and dorsal to the tonsils may not effectively select patients in whom bone decompression alone is sufficient (34). Nair et al, (46) proposed a grading system to evaluate the correlation between pre and post operative clinical and radiological parameters and to prognosticate its outcome.

**Radiological Scoring System (46)**

<table>
<thead>
<tr>
<th>Cyst : Cord</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>0.76 – 1</td>
<td>4</td>
</tr>
<tr>
<td>0.51 – 0.75</td>
<td>3</td>
</tr>
<tr>
<td>0.26 – 0.50</td>
<td>2</td>
</tr>
<tr>
<td>0.01 -0.25</td>
<td>1</td>
</tr>
<tr>
<td>0.00</td>
<td>0</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Cord : Canal</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>0.91 – 1</td>
<td>4</td>
</tr>
<tr>
<td>0.81 -0.9</td>
<td>3</td>
</tr>
<tr>
<td>0.71 – 0.8</td>
<td>2</td>
</tr>
<tr>
<td>0.61 – 0.7</td>
<td>1</td>
</tr>
<tr>
<td>&lt; 0.6</td>
<td>0</td>
</tr>
</tbody>
</table>

**Radiological Grades (46)**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Grade 0</td>
<td>No Cyst</td>
<td>0</td>
</tr>
<tr>
<td>Grade I</td>
<td>Mild</td>
<td>1-4</td>
</tr>
<tr>
<td>Grade II</td>
<td>Severe</td>
<td>5-8</td>
</tr>
</tbody>
</table>
PATHOLOGY:

Koga et al. (31) were the first to report Purkinje cell loss and reactive gliosis in the resected cerebellar tonsils of patients with CM-I. Their series consisted of 4 adult patients, all of whom had syringomyelia, and described similar histological findings in each of the cases. Pueyrredon and colleagues (54) subsequently confirmed these initial findings in a series of 43 pediatric patients with CM-I in whom the tonsils were resected at the time of surgery. They reported histological alterations in 38 of 43 samples, with the remaining 5 specimens showing no evidence of abnormality. The most frequent finding in the series was Purkinje cell loss (present in 32 specimens), followed by gliosis. Additional notable findings included internal granular cell layer loss, focal degenerative changes, and anoxic neuronal changes. The observed histological changes most likely represented a focal phenomenon secondary to specific conditions, namely local ischemia and trauma, resulting from neural tissue abnormally constrained within the narrow confines of the craniocervical junction. Purkinje cells have been shown to be especially vulnerable to ischemic insult (66). Experimental data suggest that 2 specific properties of Purkinje cells—their reduced capacity to sequester glutamate and an inability to generate energy during periods of relative
anoxia—render them particularly susceptible to ischemic death. Trapped within the foramen magnum and upper cervical canal, the distal tonsils could potentially undergo chronic ischemic changes resulting from focal constriction of arterial afferents and/or compromise of venous drainage. Local mechanical trauma to the tonsils as they are subjected to the constant cephalocaudal pistoning of CSF pulsations through the craniocervical junction likely also plays a role in the formation of chronic degenerative changes. Alteration in the local microenvironment resulting from compromise of the blood-brain barrier, excitotoxicity, and elaboration of neurotoxic molecules from local reactive microglia have all been speculated to contribute to the observed histological changes. Formation of a cyst at the distal tip of a cerebellar tonsil lying constrained within the cervical spinal canal, represents the culmination of extensive degenerative changes including gliosis and neuronal loss. These histopathological processes are likely the result of focal, chronic ischemic changes as well as repeated local mechanical trauma.
MANAGEMENT:

The indications for surgery are (37):

1) Patients with radiographic evidence of cerebellar ectopia and neurologic deficits attributable to Chiari malformation (such as - cerebellar signs, cranial nerve deficit, motor and/or sensory signs), hydrocephalus, progressive scoliosis, or syringomyelia.

2) The indication for those whose only symptom is headache is less clear. The cine – MRI is to document the obstruction of CSF flow at CVJ. Presence of obstruction to CSF flow at CVJ can be considered for surgical decompression.

The goals of surgery are to

1) Decompress the brainstem

2) Restore pulsatile flow of CSF at the cervicomedullary junction.

Decompression of the brainstem is performed via the removal of the suboccipital bone and the dorsal arch of C1 and/or C2. After bony removal, the intradural contents are explored. Any adhesions observed are lysed, thus restoring the egress of CSF from the fourth ventricle and across the cervicomedullary junction. The placement of a patulous dural graft also aids in the flow of CSF across the junction.
**Surgical steps for foramen magnum decompression:**

**Position:** Patients are intubated in the supine position. An arterial line and indwelling Foley catheter are placed. Antibiotics with a spectrum against common skin flora are given at least 30 minutes prior to skin incision. After intubation, a 3-point Mayfield head-holder is placed. The patient is then turned into the prone position onto blanket rolls (37). The head is fixed in a slightly flexed posture. This facilitates exposure and decompression. Flexion is avoided in the presence of associated bony CV junction anomalies like basilar invagination. The head of the table is kept above heart level. All pressure points are carefully padded. With the arms at the patient’s side, the shoulders may be taped back with care taken to avoid injury to the brachial plexus.

![Figure 6](image)
Exposure:

The incision is begun immediately below the inion and is carried to and past the spinous process of C2. The incision should not be carried over the inion because pressure over this bony prominence when the patient is supine may increase the incidence of wound dehiscence. After the skin is incised, Bovie cautery is used to carry the incision through the subcutaneous fat to the fascia. The muscles are divided in the midline fascial sulcus. Care should be taken not to Bovie into the muscle itself. This may lead to muscle shrinkage and increased postoperative pain. The dissection is carried down to the sub occipital bone and over the dorsal tubercle of C1 and the C2 spinous process. The muscles are first stripped laterally off the sub occipital bone. This may be done with a periostial elevator or Bovie cautery. A “V” of muscle insertion should be left in the midline immediately below the inion. The muscle dissection continues until about 3cm of bone is exposed on each side of midline. The foramen magnum is fully identified. Care should be made when using the Bovie near this region; the authors prefer to use an angled curette to clear any muscle off the edge of the foramen magnum. Angled cerebellar retractors are used to maintain the exposure. Attention should then be placed on the C1 lamina. The Bovie may be used for the
most medial dissection, but sharp dissection should be used for the lateral aspect. This decreases the risk of vascular injury (eg, vertebral artery). The vertebral venous plexus may be encountered during this part of the operation. Bleeding may be profuse. If a definite vessel is visualized it may be controlled with bipolar cautery, but the bleeding is usually controlled with a piece of thrombin-soaked Gelfoam and gentle pressure. If the cerebellar ectopia is above the C2 lamina, the authors do not strip the muscles off C2, but if the preoperative MRI demonstrates herniation below C2, the spinous process and lamina are exposed for laminectomy or partial laminectomy. A second cerebellar retractor is used to maintain the spinal exposure.

![Figure. 7](image-url)
**Bone Removal:**

A craniectomy should be planned about 2cm above the foramen magnum in children and 3cm in adults. Too generous of a craniectomy will encourage cerebellar sagging or ptosis. The appropriate extent of bone removal assessed by the sagittal MRI and removing enough occipital bone to decompress the tonsils. The craniectomy is performed with a high-speed drill and cutting burr. The drill is used to thin the bone first over the cerebellar hemispheres. Next, the thick bony keel should be thinned. This bone is often vascular and bleeding is easily controlled with bone wax. The drilling should proceed rostral to caudal. Drilling near the foramen magnum is performed last. Once the bone is adequately thinned, a curette is used to expose the dura mater over one of the cerebellar hemispheres. Leksell or large Kerrison ronguers are then used to remove the remaining bone, thus fully exposing the dura over the cerebellar hemispheres and the cervicomedullary junction. The decompression should be performed until just medial to the occipital condyles and the ring of the foramen magnum begins to turn ventrally. Venous bleeding may be encountered during this widening, but may be easily controlled with thrombin-soaked Gelfoam. A C1 laminectomy is performed next, either with the aid of the drill or the Leksell ronguer.
The laminectomy should only be as wide as the underlying dura mater. An excessively wide decompression places the vertebral artery at risk. If the tonsillar herniation is below the C1 lamina, partial C2 laminectomy will also be required.

Dural Opening:

There is often a tight ring of redundant tissue at the cervicomedullary junction. This should be incised and reflected laterally before the dural
opening. The dura mater is opened in a “Y” fashion with a 15 scalpel blade, beginning in the midline over the cervical spinal cord. Small scalpel strokes are made until the arachnoid is encountered but not violated. A dural guide is then placed subdurally, but above the arachnoid, and the dura is opened with the blade cutting immediately upon the advancing dural guide. As the cerebellar dura is approached at the cervicomedullary junction, bleeding from the circular or occipital sinuses may be encountered. If bleeding is encountered, it should be controlled with bipolar cautery or via the placement of small metal clips. The final flap of dura is sutured rostral to fully expose the cerebellum, lower brain stem, and upper cervical spinal cord. The arachnoid should be intact.

Figure. 9
Intradural Exploration:

Some surgeons do not open the arachnoid and proceed immediately to the placement of the dural graft (65). Some examine the flow of CSF and movement of the cerebellar tonsils with ultrasound and do not open the dura mater or arachnoid if adequate movement and flow of CSF is established after the craniectomy (50).

Jorg Klekamp et al, (25) intraoperative findings, the arachnoid pathology was graded according to the following criteria:

Grade 0 = no arachnoid pathology detectable

Grade 1 = slight arachnoid adhesions to cerebellum or spinal cord, arachnoid translucent
Grade 2 = severe arachnoid scarring, arachnoid not translucent, dense adhesions to cerebellum, brainstem, or spinal cord.

If the arachnoid is to be opened, it is incised sharply and is attached to the dura mater with suture or metalclips. Using loupe magnification or the operating microscope, the tonsils and the foramen of Magendie are examined. A number of points need to be stressed in this respect. Keeping the subarachnoid space clear from any contamination with blood is mandatory to limit postoperative arachnoid scar formation. Arachnoid dissection should be restricted to the midline, avoiding cranial nerves and perforating vessels using sharp dissection only to avoid any tearing on these structures (25). Often the tonsils are adherent to each other or the brainstem by thickened arachnoid. A Penfield 4 is used to separate the 2 tonsils from each other and identify PICA. The goal is to separate the tonsils to permit the egress of CSF from the fourth ventricle. Care should be taken to not damage PICA or try to dissect adherent tonsils from the brainstem. If the tonsils are not easily separated, bipolar cautery may be used to shrink the arachnoid and hence the tonsils, which should permit the flow of CSF from the fourth ventricle. If the scarring is too dense, a more aggressive strategy is used.
The tonsils may be removed via an endopial resection. PICA should be unequivocally identified prior to the subpial resection.

**Dural Grafting:**

Multiple choices of grafting materials are available. Autologous choices include pericranium, ligamentum nuchae, and fascia lata. Non-autologous options include cadaveric dura, lypholized dura or fascia, bovine pericardium, human pericardium, and Gor-Tex. Autologous graft material is preferred whenever possible. The wound should be copiously irrigated prior to the placement of the final sutures, to ensure the removal of blood products and verify that there is not any active bleeding. The anesthesiologist should perform a Valsalva maneuver to check the integrity of the suture line. If any sites of leaking are visualized, simple sutures are placed. A small piece of muscle may also be sutured at the site of a leak.

Figure. 11
Muscle and Skin Closure:

The muscle should be closed in layers with absorbable suture. The fascia is closed and reattached to the muscle and fascia along the ligamentum nuchae. The midline is attached to the cuff of muscle that was spared during the initial exposure. The dermis is closed and the skin closed with sutures or staples.

POSTOPERATIVE CARE:

All patients are closely observed for 24 hours postoperatively. The patients should have full cardiovascular and respiratory monitoring and close observation for neurologic changes. On postoperative day 1 patients are fully mobilized. Their diet is advanced, but any signs of aspiration should be carefully sought. A soft collar may be prescribed for comfort only. Patients return to clinic at 6 weeks and 3 months with an MRI of the craniovertebral junction and cine-MRI flow study. This is to ensure adequate flow of CSF at the cervicomedullary junction and a decrease in the size of any syrinx that was present preoperatively. It serves as a baseline study for future care.
POSTOPERATIVE COMPLICATIONS

Early Complications:

- CSF leak and/or pseudomeningocele
- Meningitis
  - Infective
  - Chemical
- Hematoma

Late Complications:

- New or enlarging syrinx
- Obstruction of CSF flow across the cervicomedullary junction due to scarring
- Cerebellar ptosis.

Early Complications

By far the most common early complication is CSF leak and/or pseudomeningocele. Meticulous surgical technique should minimize the incidence of this complication. CSF leak should first be treated by over sewing the wound. If this fails, placement of a lumbar subarachnoid drain is appropriate. If the lumbar drain fails, re-exploration may be necessary.
Meningitis has been reported to occur following Chiari decompression. Contamination usually occurs at the time of surgery, and may be minimized by attention to detail (such as preoperative antibiotics, careful skin preparation, etc.). If non-autologous graft material was used, removal of the graft may be required if the infection cannot be cleared with antibiotics alone.

Chemical meningitis also frequently occurs following this procedure. It may be due to blood in the CSF or immune reaction to non-autologous graft material. Diagnosis is made by spinal tap with negative cultures. Often eosinophils are present in the CSF.

Patients may have temporary or permanent lower cranial nerve paresis. This may lead to aspiration or minor problems with respiration. These complications may be minimized by not trying to aggressively dissect the cerebellar tonsils off the brain stem. If symptoms are severe, tracheotomy and/or gastrostomy tubes may be required.

The most feared complication is hematoma. Clinical deterioration may be rapid and is the main reason for close neurologic observation postoperatively. If clinical deterioration is observed following surgery, the patient should be taken for emergent CT scan and then to the OR for hematoma evacuation. If rapid deterioration occurs, emergent
decompression at the bedside (with closure in the operating room) may rarely be required.

**Late Complications**

Clinical deterioration may occur after initial improvement following surgery.

Risk Factors of a Neurological Recurrence (25)

- Arachnoid pathology
- Less experienced surgeon
- Arachnoid not opened
- Basilar invagination

An MRI of the brain and cervical spinal cord and a cine-MRI flow study are indicated under these circumstances. Common etiologies include new or enlarging syrinx, obstruction of CSF flow across the cervicomedullary junction due to scarring, and cerebellar ptosis. All 3 may require reoperation and exploration of the prior decompression. A “shelf” of methylmethacrylate may be fashioned in the management of cerebellar ptosis. An anatomy-specific re-decompression may be indicated in other cases, with the goal being the establishment and maintenance of normal or physiological unobstructed pulsatile flow of CSF in the cervicomedullary junction region.
OUTCOME

Patient prognosis and long-term response to surgery remain highly variable. Poor outcome is frequently observed in patients with signs or symptoms suggestive of syringohydromyelia, although less common, atrophy, ataxia, and nystagmus suggest a similarly poor prognosis. Headache specifically and pain in general appear to respond best. Weakness in the absence of atrophy tends to respond well, whereas weakness in the presence of atrophy is uniformly unresponsive to surgery. Scoliosis responds reasonably well to surgery. Sensory loss has been widely recognized as largely unresponsive to surgery. Patients with physiologically significant symptoms and correlative MRI and cine-MRI flow findings, approximately 90% will have improvement or stabilization of their symptoms (16). Most patients with an associated syrinx are observed to have a decrease in the size of the syrinx within the first 3 months following surgery (15). Dyste G N et al (15) reported 81.5 % resolution of pain, 21 % preoperative weakness return to normal strength, no patient with muscle atrophy return to normal strength.
J. C. Alazate et al (27) Pain was more likely to resolve than sensory and motor deficits after decompressive surgery. Jorg Klekemp et al (25) reported 73.6 % of patients improved after 3 months, most profound effect seen in occipital pain which almost always improved after surgery. K. S. Paul et al (28) – most favorable long-term results were seen in patients with the cerebellar syndrome 87% of these showed improvement and none experienced late deterioration. Patients presenting with foramen magnum compression, 81% showed early
improvement but one-third of these later deteriorated. Patients with a central cord syndrome also showed encouraging early postoperative improvement (84%), but one-quarter of these subsequently deteriorated.
AIM OF THE STUDY

To retrospectively study children (≤ 18 years) with chiari I malformation treated surgically at our institute and analyze the long-term outcome of treatment.
MATERIALS AND METHODS

All patients who were surgically treated at the Department of Neurosurgery, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram, with the diagnosis of chiari type I malformation with age \( \leq 18 \) years, their charts retrospectively reviewed for the period of January 1999 to June 2011. Data were collected regarding demographics, clinical symptoms and signs at the time of admission, Magnetic resonance imaging (MRI) features and postoperative complications. Post-operative follow up at 6 weeks, 6, 12, 24 months was done to evaluate resolution of signs and symptoms, and imaging features. Our results were compared to data in previously mentioned literature.
RESULTS

The results obtained from the study are expressed in the following section in tabular format. Numerical, continuous data are expressed with mean.

Demographics:

There were 21 girls (42%) and 29 boys (58%), with female to male ratio of 1 : 1.4. The average age of the patients was 13.8 years, with a range of 1.5 to 18 years.

Figure 13: Sex Distribution
Clinical characteristics:

The 3 common presentations were headache/neck pain 46 % (n = 23), upper limb pain/numbness/weakness 46 % (n = 23) and scoliosis in 24 % (n = 12). Other presentations were gait ataxia 12 %, lower limb weakness and hyperreflexia 12% and features of lower cranial nerve deficit 18 % (nasal speech 8 %, dysphagia 6 % and hoarseness of voice 4 %), dyspnea, facial numbness, chronic emesis, chronic hiccups, drop attack, urinary incontinence and hearing loss.
Signs and Symptoms

- Head/Neck pain: 23
- UE Pain/weakness/Numbness: 23
- Lower C. N. Dysfunction: 9
- Scoliosis: 1
- Ataxia: 6
- Dyspnea: 3
- Facial Numbness: 2
- Chronic emesis: 2
- Chronic hiccups: 1
- Drop Attack: 1
- Urinary Incontinance: 1
- Hearing Loss: 1

Figure 15: Clinical Characteristics
Magnetic resonance imaging:

All the patients were evaluated with magnetic resonance imaging to look for Tonsillar descent, presence and extent of syrinx, scoliosis. Tonsillar descent was present in all the patients, of which in 31 (range: 5 – 26 mm; mean 14.5mm) the extent of tonsillar herniation was measured and mentioned in case records. In the rest 19 patients, only the relation of the inferior border of cerebellar tonsils to C1, C2 and C3 was documented.

![Tonsillar descent - mm](image)

**Figure. 16**

Syrinx was present in 80 % (n = 40). Most commonly involved segment in syrinx was cervico-dorsal 77.5 %, only cervical in 17.5 %, only thoracic segment 2.5 %, holocord syrinx 2.5 %.
Scoliosis was present in 24 %. Scoliosis was more common toward right side 58.3 % and toward left side 41.6 %. Scoliosis observed more commonly in male patients 58.3 %, in female patients 41.6 %. Scoliosis most commonly involved cervicodorsal region 75 %, followed by cervical and thoracic region equally 12.5 % each.

**Surgical procedure and Intraoperative findings:**

Of the 50 patients with chiari I malformation, 94.00 % (n = 47) underwent foramen magnum decompression with lax duroplasty, (n = 1) treated with foramen magnum decompression with lax duroplasty and
Tonsillar resection (left tonsil was gliotic), \( n = 1 \) treated with foramen magnum decompression with lax duroplasty with C2 – C3 posterior fusion (Subluxation of C2 - C3), \( n = 1 \) treated with foramen magnum decompression with lax duroplasty with occipito-cervical fusion with contour rod (reducible atlantoaxial dislocation). Indication of tonsillar resection was that during intraoperative left tonsil was gliotic and more elongated. Indication for C2 – C3 posterior fusion was subluxation of C2 – C3 and indication for occipito-cervical fusion was reducible atlantoaxial dislocation.

![Figure. 18 : Surgery](image)

**Figure. 18 : Surgery**
Patients posterior cranial fossa decompression with lax duroplasty with removal of the posterior arch of C-1 with lax duroplasty using autologous pericranium. The posterior arch of C-1 was bifid in 10% of patients. After removing the posterior arch of C-1, a dense constrictive band causing compression was observed in 40% of patients. The dura mater of the craniocervical junction was thickened, based on subjective assessment, in 30% of patients. At operation, 25% of patients with syringomyelia were found to have an arachnoid veil occluding the fourth ventricular outlet, and in each case the veil was transected. Unilateral tonsillar resection was performed in 1 patient. Other anomalies noted at operation included thickened midline keels (internal occipital crests), lateral narrowing of the foramen magnum, arachnoid adhesions within the fourth ventricle, and a low-lying transverse sinus

Outcomes:

At 6 weeks: At 6 weeks follow up there was no improvement in 28%. Lower cranial nerve dysfunction improved in 33%, headache/neck/back pain improved in 56.5%, upper extremity pain/weakness/numbness improved in 78.3%, ataxia improved in 66.7%, lower limb weakness/hyperreflexia improved in 66.7%. 4% patients developed intermittent diplopia at 6 weeks follow up.
At 6 months: At 6 month follow up headache/neck/back pain improved in 5 more patients, at 6 months, headache/neck/back pain improved in 78.3% patients. Upper extremity pain/weakness/numbness improved in 3 more patients, at 6 months it improved in 91.3%. 6 patients with persistent lower cranial nerve dysfunction at 6 months lost follow up. No other symptoms were improved.

At 12 month: At 12 month follow up total 3 patients were lost to follow up and in 1 patient lower limb spasticity improved; at 12 month 83.3% lower limb weakness/hyperreflexia improved. 1 patients developed headache which was relieved earlier (managed on conservative treatment).

![Figure 19: Clinical Results at 12 Month follow up](image)
**At 24 months:** At 2 year follow up out of the 38 patients who were available, 4 patients developed new upper limb pain/weakness/numbness. Others continued to remain asymptomatic.

**Last follow up:** Mean duration of follow up 45.5 months (Minimum 3 months, maximum 155 months). At the time of last follow up 7 patients - no improvement of symptoms and 4 patients developed new symptoms of upper limb pain/weakness/numbness. 2 patients referred to orthopaedician for scoliosis surgery. 1 patient referred to psychiatrist for multiple somatic complaints.

![Figure 20: Clinical Results at last follow up](image)
At last follow up, lower cranial nerve dysfunction improved in 33 %. Headache/neck/back pain improved in 69.6 %, upper extremity pain/weakness/numbness improved in 73.9 %, ataxia improved in 66.7 %, lower limb weakness/hyperreflexia improved in 83.3 %. At follow up MRI for patients with syrinx was available for 75 % patients, and not available for 25 % patients. Syrinx was diminished in size or resolved in 66.3 % patients and remaining same for 36.7 %.

![Figure, 21: follow up MRI (syrinx)](image)

Complications: Out of 50 patients tracheostomy done in 3 patients, 2 patients due to lower cranial nerve paresis and in 1 patient for elective ventilation due to poor respiratory reserve preoperatively. All the 3
patients were referred to ENT surgeon for evaluation and decannulation of tracheostomy. All the 3 patients were doing well at 6 weeks follow up. None of the patient developed wound infection. 1 patient developed left PICA territory infarct with was managed with antiedema measures, 1 patient developed intermittent diplopia in postoperative period. 3 patients developed fever which was managed with antibiotics (No CSF study was done).

![Complications - Number Of Patients](image)

**Figure, 22: Complications**
Case 1:

A 16 year old girl presented with a 4-month history of numbness in her right arm. A neurological examination revealed decreased pain and temperature sensation in right C5-T1 dermatome. Tone in both lower limbs grade I increased with right plantar was extensor. The remainder of the examination was normal. A magnetic resonance imaging (MRI) scan showed peg like tonsillar herniation up to upper border of C2 with syrinx from C3-D5. She underwent foramen magnum decompression and lax duroplasty. Postoperative imaging showed significant resolution of syrinx and her symptoms improved.
Figure 23: Pre-operative MRI Imaging showing Peg like downward displacement of cerebellar tonsils with syrinx C4-T5 (Lower margin of syrinx not seen)
Case 2:

A 17 year old boy presented with a 9-month history of neck and suboccipital pain which aggravated by cough. Neurological examination revealed absent bilateral supinator and triceps jerk with left C2 hypoesthesia and loss of pain and temperature in left T6-T10 dermatome. The rest of the examination was normal.

A magnetic resonance imaging (MRI) scan showed peg like tonsillar herniation 15 mm below foramen magnum with syrinx from CVJ-D11. He underwent foramen magnum decompression and lax duroplasty. Postoperative imaging showed significant resolution of syrinx and his symptoms improved.
Figure 25: Pre-operative MRI Imaging showing Peg like downward displacement of cerebellar tonsils 15 mm below foramen magnum with syrinx CVJ-T11 (Lower margin of syrinx not seen)
Figure, 26; Post-Operative changes with Significant Reduction of Syrinx
DISCUSSION

Chiari malformation Type I is characterized by caudal descent of cerebellar tonsils and may or may not be associated with the presence of a syrinx, a degree of medullary descent and buckling of the lower medulla may also be present. A tonsillar herniation of more than 5 mm is widely considered pathological in adults. In children, cerebellar and neocortical growth causes a physiological herniation of the cerebellar tonsils, whereas in old age atrophy of the brain may lead to tonsillar ascent. In doubtful cases, cardiac gated cine-MRI is very helpful to demonstrate a CSF flow obstruction as an indicator of a clinically relevant herniation. Symptoms of Chiari malformations are related to age: in infancy, signs of brainstem compression predominate with apnea spells, cyanosis attacks, and swallowing problems, whereas in later childhood scoliosis becomes the most common presenting sign. The typical clinical features of occipital headache, gait ataxia, sensory disturbances, and motor weakness are uncommon in children and observed predominantly in adults.
DEMOPGRAPHICS:

The average age of the patients in the study 13.85 years, with a range of 1.5 to 18 years, female to male ratio is 1:1.4. R. S. Tubbs et al (59) between 1989 and 2010, 500 patients underwent surgery for treatment of chiari malformation type I. The age of the 500 patients in the study group ranged from 2 months to 20 years (mean 11 years), female to male ratio 1 :1.18. M. J. McGirt (61) present data of 130 pediatric patients with Chiari I malformations whom underwent surgery in 23 years of surgical experience. Patients age ranged from 2 months to 20 years (mean 11 years), female to male ratio 1 : 1.13. A. H. Menezes et al (15) between 1975 and 1985, 50 symptomatic patients with Chiari malformation type I underwent surgical treatment. The pediatric group comprised 16 patients below the age of 20 years, with the remaining 34 patients forming the adult group. Among the pediatric patients the average age was 11 years (range 1 to 19 years). Our study is in concordance with given study by R. S. Tubbs et al. and M. J. McGirt et al in view of sex distribution. Average age of patients is slightly higher in our study (13.85 years) as compared to previous studies (11 years).
CLINICAL CHARACTERISTICS:

In present study 46% patients presented with headache/neck pain. In a series by R. S. Tubbs et al (59) headache/neck pain was the presentation in 40%. M. J. McGirt et al, (61) patients presented with headache/neck pain were 42.3%. A. H. Menezes et al (15) in paediatric group ≈ 40% had pain as presentation. Carmine Mottolese et al (10) 40.00% of children with chiari I malformation presented with headache in group A and 53.00% of children in group B. Presentation as headache is in concordance with with earlier published studies.

In present study the upper limb pain/numbness/weakness is the presentation in 46%. In a series by R. S. Tubbs et al. (59) upper limb pain/numbness/weakness was present in 8.2% of patients. M. J. McGirt et al, (61) 16.9% patients presented with upper limb pain/numbness/weakness. A. H. Menezes et al (15) in pediatric group ≈ 80% had motor signs/symptoms. Carmine Mottolese et al (10) 15% of children presented with motor deficit and 16% with trouble in sensibility group A. Presentation as upper limb pain/numbness/weakness is more in present study as compared to earlier studies.
In present study 24% of patients had scoliosis. In a series by R. S. Tubbs et al. (59) scoliosis was present in 18%. M. J. McGirt et al (61) 17.7% of patients had scoliosis. A. H. Menezes et al (15) in pediatric group, scoliosis was present in ≈ 50% of patients. Carmine Mottolese et al (10) 16.5% of children with chiari I malformation had scoliosis in group A and 17% of children in group B. In the present study scoliosis is more common toward the right side (58.3%) as compared to left side (41.7%). Scoliosis is more common in males (58.3%) as compared to females (41.7%).

In present study 12% patients presented with lower limb weakness and hyperreflexia. In a series by R. S. Tubbs et al. (59) lower limb weakness and hyperreflexia was the present in 2.4%. M. J. McGirt et al, (61) lower limb weakness and hyperreflexia was present in 5.4%. In present series 8% patients have quadriparesis (4 patients common in both upper limb weakness and lower limb weakness). A. H. Menezes et al (15) 12% were presented with quadriparesis.

In present study 12 patients presented with ataxia. In a series by R. S. Tubbs et al. (59) ataxia was the present in 3.8%. M. J. McGirt et al, (61) patients ataxia was present in 9.2%. A. H. Menezes et al (15) in paediatric group ≈ 20% had ataxia. Our study is in concordance with
given study by M. J. McGirt et al. Presentation as ataxia varied significantly in earlier series.

In present study features of lower cranial nerve deficit - nasal speech, dysphagia and hoarseness of voice observed in 18%. R. S. Tubbs et al. (59) reported lower cranial nerve deficit 8.8%. M. J. McGirt et al, (61) patients with lower cranial nerve deficit were 33%. Other signs and symptoms are compared in Table 1.
<table>
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<th>Sign/symptoms</th>
<th>M. J. McGirt et al</th>
<th>R. S. Tubbs et al</th>
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<td>19(3.8%)</td>
<td>6(12%)</td>
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<td>Nasal Speech</td>
<td>15(11.53%)</td>
<td>15(3%)</td>
<td>4(8%)</td>
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<td>Drop Attack</td>
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<td>Hearing loss</td>
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Magnetic resonance imaging:

Tonsillar descent was present in all the patients, of which in 31 the extent of tonsillar herniation was measured and mentioned in case records. In the rest 19 patients, only the relation of the inferior border of cerebellar tonsils to C1, C2 and C3 was documented.

Tonsillar descent:

5 – 10 mm – 11 patients,
11-20 mm – 14 patients,
> 20 mm - 6 patients

Figure 27: Comparison with Earlier studies
In other 19 patients lower limit of tonsil was between foramen magnum and C1 in 4 patients, up to C1 level in 10 patients and up to C2 level in 5 patients. In a series by R. S. Tubbs et al. (59) the inferior border of the lowest cerebellar tonsil between the foramen magnum and C-1 in 22 % patients, at C-1 in 37.4 % patients, at C-2 in 39.4 % and at C-3 in 1.2 %. M. J. McGirt et al, (61) the inferior border of the lowest cerebellar tonsil to be situated between the foramen magnum and C-1 in 20 % patients, at the level of C-1 in 46.9 % patients, at the level of C-2 in 32.3 % patients and at the level of C-3 in 0.7 % patients.

In present study 80 % of patients had syringomyelia with most commonly involved segment in spinal cord was cervico-dorsal 77.5 %, only cervical in 17.5 %, only thoracic segment 2.5 %, holocord syrinx 2.5 %. In a series by R. S. Tubbs et al. (59) syringomyelia was present in 57 % patients. Syringomyelia involving holocord was most common seen in 39.3 %, followed by cervicothoracic in 24.9 %, cervical in 15.4 %, thoracic in 14.7 %, lumbar in 3.5 % and syringobulbia in 2.1 %. M. J. McGirt et al (61) syringes were present in was present in 58 % patients. Syringomyelia involving holocord was most common seen in 44 %, followed by cervical in 21.3 %, thoracic in 16 %, cervicothoracic in 12 %, syringobulbia in 4 % and lumbar in 2.7 %. The incidence of
syrinx in pediatric patients in our study (80%) was higher than reported studies (50 – 76%). The most commonly involved segments in present study are cervicothoracic followed by cervical. In earlier studies syringomyelia was present in holocord followed by cervicothoracic. Syringobulbia and syrinx involving the lumbar region are not seen in present study.

**Surgical procedure:**

In present study 94% underwent foramen magnum decompression with lax duroplasty, (n = 1) treated with foramen magnum decompression with lax duroplasty and Tonsillar resection (left tonsil was gliotic), (n = 1) treated with foramen magnum decompression with lax duroplasty with C2 – C3 posterior fusion (Subluxation of C2 - C3), (n = 1) treated with foramen magnum decompression with lax duroplasty with occipito-cervical fusion with contour rod (Reducible atlantoaxial dislocation). In present study pericranium was used as graft for duroplasty in all cases. In a series by R. S. Tubbs et al. (59) all patients underwent except 1 underwent foramen magnum decompression and lax duroplasty at first operation with cadaveric pericardium in 8% of cases, autologous pericranium 91%, nuchal ligament 0.5%, and posterior atlantooccipital membrane 0.5%. 1 underwent lax duroplasty at second operation due to persistent
symptoms. Posterior element of C-2 removed in 0.4% patients, tonsillar coagulation in 9.8% patients and craniospinal fusion in 2.4%. M. J. McGirt et al. (61) all patients underwent except 1 underwent foramen magnum decompression and lax duroplasty at first operation. 1 underwent lax duroplasty at second operation due to persistent symptoms. Posterior element of C-2 removed in 0.8% patients, tonsillar coagulation in 16.9%. Cervical instability was noted in one patient preoperatively with no clinical symptoms indicative of cervical instability in follow up. Foramen magnum decompression with lax duroplasty is most commonly done surgery in all studies with tonsillar coagulation more in earlier studies.

**Complications:**

In the present study 6% patients underwent tracheostomy, 2 patients due to lower cranial nerve paresis and in 1 patient for elective ventilation due to poor respiratory reserve preoperatively. None of the patient developed wound infection. 2% patient developed left PICA territory infarct with was managed with conservatively, 1 patient developed intermittent diplopia in postoperative period. 6% patients developed fever which was managed with antibiotics (No CSF study was done). In a series by R. S. Tubbs et al. (59) complication occurred
in 2.4 %. 0.8% developed subdural extra axial collection which was managed with external ventricular drainage. 0.8 % developed severe life-threatening signs of brainstem compression following surgery managed with a transoral odontoidectomy and occipitocervical fusion. 0.2% developed bacterial meningitis (treated with antibiotics) and 0.2 % developed CSF leak (managed with shunt placement). No arterial injury or any death. M. J. McGirt et al (61) complication occurred in 2.3 %. 1.5 % developed subdural extra axial collection which was managed with external ventricular drainage. 0.8 % developed severe life-threatening signs of brainstem compression following surgery managed with a transoral odontoidectomy and occipitocervical fusion. No infection, CSF leak, arterial injury or any death.

**Outcomes:**

Mean duration of follow up in present study is 45.52 months (minimum – 3 months, maximum 155 months). At last follow up, headache/neck/back pain improved in 69.6 %. Upper extremity pain/weakness/numbness improved in 73.9 %, ataxia improved in 66.7 %, lower limb weakness/hyperreflexia improved in 83.33 %, lower cranial nerve dysfunctions were improved in 33 %. At follow up MRI for patients with syrinx was available for 75.00 %(n = 30/40) patients,
and not available for 25.00 % (n = 10/40) patients. Syrinx was diminished in size or resolved in 66.3 % (n = 19/30) patients and remaining same for 36.7 % (n = 11/30). R. S. Tubbs et al. (59) posterior fossa decompression relieves symptoms of a syrinx in 80% of cases and 83 % relieve in pre-operative signs and symptoms. M. J. McGirt et al, (61) posterior fossa decompression relieves headache/neck/back pain in 80% of cases and 83 % relieve in pre-operative signs and symptoms. Symptomatic improvement is less in present study as compared to earlier studies.
CONCLUSIONS

Chiari malformation Type I represents a complex anatomical and clinical challenge. The incidence of syrinx in pediatric patients in our study (80 %) was higher than reported studies (50 – 76%). Patients with a symptomatic Chiari malformation and/or a distended syrinx cavity are ideal candidates for surgery where the main goal of surgery is to arrest the progression of neurological deficits. The role of surgery in patients with asymptomatic chiari malformation continues to be debatable. Foramen magnum decompression with lax duroplasty is the surgical procedure of choice although the decision to perform additional steps depends on the operating surgeon. The extent of decompression should be sufficient enough for CSF flow across the foramen magnum. The risk of complications from duroplasty was minimal in current series. On long term follow up, no problems related to cervical spine instability was noted following removal of posterior arch of C1. Proper patient selection is necessary for achieving appropriate postoperative results.
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PROFORMA FOR PAEDIATRIC (≤ 18 YRS) CHIARI 1 MALFORMATION

1. Name: Age: Sex:

2. Presenting Complaints:

- Oropharyngeal dysfunction
  - Dysphagia Y/N
  - Hoarseness Y/N
  - Nasal speech Y/N
- Headache/Neck/Back pain Y/N
- Scoliosis Y/N
- Drop Attacks Y/N
- Upper limb pain/weakness/Numbness Y/N
- Lower limb hyperreflexia/weakness Y/N
- Facial numbness Y/N
- Gait ataxia Y/N
- Dyspnea Y/N
- Chronic Hiccups Y/N
- Chronic emesis Y/N
- Urinary Incontinance Y/N
- Hearing Loss Y/N

3. Imaging

MRI

Tonsillar descent Y/N cms:

Syrinx Y/N Level:

Scoliosis Y/N

4. Surgery

- FMD Y/N
- FMD with lax duraplasty Y/N
- FMD, lax duraplasty and tonsillar resection Y/N
Any other surgery

5. Complications

- Lower cranial nerve palsy
- Tracheostomy
- Poor respiration and elective Ventilation
- Wound infection
- Others:

6. Follow up

- 6 weeks: Resolution of symptoms: Y/N
  If yes, which symptoms resolved:
  - 6 months:
  - 12 months:
  - 24 months:

7. Follow up MRI:

Resolution of syrinx: Y/N

8. Whether following resolved?

- Headache: Y/N
- Scoliosis Y/N
- Oropharyngeal dysfunction Y/N
- Syrinx Y/N

9. Recurrence: Y/N

If yes, details: