

# Original article

## Neural endoscopic assisted micro-invasive management of Chiari I malformation

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**Keywords:** *Chiari I malformation; syringomyelia; neural endoscope; posterior fossa decompression; minimally invasive neurosurgery*

**Background** In order to make posterior fossa decompression for the management of Chiari I malformation simple and less invasive while using direct visualization, a novel solely endoscopic procedure has been employed for the decompression of Chiari malformation type I. The objective of this study was to present neural endoscopic posterior fossa decompression and atlas laminectomy for Chiari type I patients.

**Methods** Twenty-one patients with Chiari type I underwent neural endoscopic posterior fossa decompression and atlas laminectomy. We described the procedure for neural endoscopic posterior fossa decompression and atlas laminectomy. All patients in this series demonstrated cerebellar tonsil herniation below the foramen magnum in addition to syringomyelia. All patients in the reviewed study underwent preoperative MRI as well as 3-month postoperative MRI. Additional follow-up MRI varied but was usually repeated 12 months to 18 months after surgery. Postoperative MRI studies were retrospectively reviewed and compared with preoperative studies.

**Results** All patients showed clinical improvements, and none had any complications. Patients with syringomyelia had symptoms entirely disappear. Eleven patients (52.4%) experienced radiographic improvement in syringomyelia (decreased size or resolution) during the follow-up period. Nine patients (42.8%) demonstrated decreased syrinx size and four (19%) demonstrated resolved syrinx. Of the 15 patients with symptomatic syringomyelia, 11 (73.3%) experienced symptomatic improvement. The median time to symptom improvement was four months after surgery. Post surgical MRI examinations indicated complete and sufficient decompression of foramen magnum region.

**Conclusions** Endoscope atlanto-occipital decompression surgery is an innovative, safe and effective surgical procedure. It has similar results compared to traditional surgery, however with the added advantages of being minimal invasive, having fewer complications, decreased influence on stability of occipital bony structure, and a faster recovery as well as reduced hospital stay and expenses.

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Amygdala cerebelli herniation, also termed Chiari malformation,<sup>1</sup> is clinically categorized into 4 types, with types I and II being the most common. Presently, common surgical treatment of amygdala cerebelli herniation and syringomyelia is through a traditional centro-posterior craniotomy approach for atlanto-occipital decompression of foramen magnum. The first line of surgical therapy at many institutions is craniovertebral decompression, usually including suboccipital craniectomy with atlas. Sometimes this may also include axis laminectomy with and without intradural exploration, reduction of the tonsils, duraplasty, and even with shunt implantation for syringomyelia. The specific surgical steps in this operation continue to undergo modification as surgeons attempt to identify the optimum procedure. In addition, postoperative complications including pseudomeningocele, meningitis, CSF leak, adhesion, cerebellar ptosis and cervical instability have brought about not only failures of the operation but also potentially severe consequences to some of the patients. Management of Chiari I malformation with less postoperative complication and recurrence continues to pose great challenges to surgeons.<sup>2,3</sup> In this study, we conclude our clinical experience for neural endoscopic

assisted micro-invasive management of Chiari I malformation

### METHODS

We retrospectively reviewed 21 consecutive pediatric patients between October 2007 and December 2008 at the Peking Union Medical College Hospital who had undergone neural endoscopic posterior fossa decompression and atlas laminectomy for CM1-associated syringomyelia. Headache was regarded as a symptom of Chiari-associated syringomyelia based strictly on its tussive nature and reproducibility in the clinic. Perioperative complications, including wound breakdown and aseptic meningitis were recorded as well.

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All patients in the reviewed study underwent preoperative MRI as well as 3-month postoperative MRI. Additional follow-up MRI varied but was usually repeated 12 months to 18 months after surgery. Postoperative MRI studies were retrospectively reviewed and compared with preoperative studies. Syringomyelia was classified as resolved if there was no evidence of residual syrinx, as decrease if the syrinx diameter was decreased by at least 20%, and as no change if the syrinx diameter was decreased by less than 20% compared with the preoperative diameter. Postoperative records were specifically evaluated for persistence of syrinx-specific symptoms; including dysesthesia, paresthesia, gait abnormalities, incontinence or weakness. Resolution of hindbrain, cranial nerve, or headache symptoms alone was not regarded as a resolution of syrinx-specific symptoms.

### Patients

All patients in this series were offered surgical decompression if they presented with symptoms consistent with CM1 (tussive headache, cervical pain, central apnea, dysphagia, aspiration, vertigo, vocal cord paralysis, motor/sensory deficits, nystagmus, ataxia, or incoordination) and demonstrated cerebellar tonsil herniation below the foramen magnum in addition to syringomyelia. We performed neural endoscopic posterior fossa decompression and atlas laminectomy in all the patients.

### Operative techniques

Patients underwent endotracheal intubation general anesthesia, ventral decubitus position with head frame fixation, top of the head elevated by 30 degrees, with natural descensus of head and ante flexion of the neck, with careful attention in maintaining stable airway conductance. The incision was normally designed above or below the hairline, above the spinous process of C2, according to the appearance of the patient's posterior cranial fossa. If the inclination angle of the squamo-occipital angle was large, the posterior cranial fossa forms a funnel like structure hence the incision should be designed further upwards. If the squamo-occipital angle was relatively planar, in order to facilitate exposure, the incision should be lower. Normally we observed the foramen magnum from an inferior position during surgery. In general we formed an alignment according to the foramen magnum and C1, using an incision length of 2.5 cm.

Fixation arms were used to fixate and stabilize the endoscope, and to assemble water and suction systems to facilitate the intra-operative cleaning of the surgical field as well as to clean the endoscope lens. After cutting the skin, under endoscopic assistance we slowly made an incision using an electrotome bovey cutting along the midline until reaching the foramen magnum. We then exposed the posterior arch of C1. The exposure site was often 3 cm above the foramen magnum, and 1.5 cm–2 cm

to the side of the foramen magnum midline, cut downward until the upper portion of C2 spinous process. This allows easy exposure, often requires cutting of the musculus rectus capitis posterior minor, preserving the musculus rectus capitis posterior major through retraction.

The exposure process required strict alignment to the midline, to prevent directional loss and confusion within a small working space. It could prevent and reduce bleeding by following the same stable midline alignment with white line anatomical landmarks during abstraction. The musculus rectus capitis posterior minor acted as anatomical landmark during abstraction into deep muscle layers, preserving the C1 posterior node of the posterior arch to act as an anatomical landmark. If the posterior node was not obvious due to abnormal development of bony structure, we would refer to the spinous process of C2 to exact and determine midline. Prior to stripping the substantia ossea, it was necessary to determine the accuracy of the midline; if the line was slightly off, it might lead to injury of the vertebral artery.

After exposure of the occipital bone and C1 posterior arch, a high speed drill was used to strip the thin substantia ossea followed by careful delicate biting using Kerrison rongeurs. Normally, we can see obvious bony structures at this point; such as, the posterior border of the foramen magnum or obvious C1 posterior arch compression or entrapment of dura mater. The range of removal of bone substantia ossea was 3 cm above the foramen magnum and about 1.5 cm–2 cm on either side. While stripping on both sides, the surgeon must again determine midline position, normally alternating between sides or use the aid of a 30-degree endoscope to enlarge the visual field. Special caution is advised when approaching the occipital condyle with specific attention to notice the vertebral artery. Due to the rich supply of para-vertebral venous plexus, venous bleeding often occurred and sufficient hemostasis could be obtained through gelatin sponge compression.

There is still debate on whether it is necessary to open the dura mater; this all depends upon intra-operative examination of whether or not there exists a bundle or girdle syndrome on the dura surface caused by entrapment and compression of bony structures. Prior to opening the dura mater, it is advised to use an ultrasound Doppler system to detect cerebral spinal fluid flow rate. If, after the decompression of bony structure there is an improved CSF flow rate, then according to experience, it is not necessary to open the dura mater. If there is thickening of the dura mater surface forming a girdle like bundle, a small Y shaped incision may be made on the superficial layer of the dura mater, and ultrasonography may be used to detect CSF flow rate. If the results indicate stenosis or obstruction, decompression of the dura should be performed through a small Y shaped incision as well as exploration and investigation of the

fourth cerebrum ventricle exits. We normally do not remove the amygdala cerebelli. It is unnecessary to suture and fix the dura after a small incision, however sometimes during the incision process, there might be bleeding of the superficial dura layers. Bleeding control may be achieved through gentle compression or electrocauterization or suture ligation. Opening of entire dura layer may lead to enlargement of the occipital sinus opening.

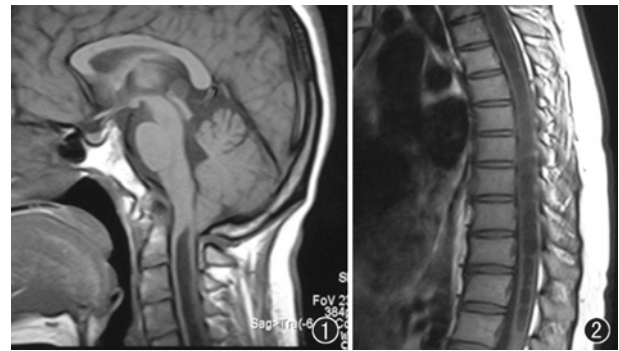
Surgical instrumentation required preparation of a narrow, deep mastoid process retractor, with a larger curved handle with an increased retracting strength, allowing for access and exposure during surgery. The abrasion burr needed to have high stability, with safe efficiency leading to reduced operation time. Also required were a multi-directional Kerrison rongeur to sustain intra-operative manipulation and handling; because of the limited surgical field and space, endoscope assistance eliminates time required for apparatus and instrument changes.

Our endoscope was produced by the Stozes Company and was required to have a fixture arm to stabilize and grip the endoscope, thus allowing the surgeon free manipulation and operation with both hands, as well as guaranteeing a stable visual field. It was simultaneously equipped with a water drainage and washing system to facilitate intra-operational cleaning and drainage of the visual field and endoscope lens.

## RESULTS

### Patient population

Twenty-one children underwent surgery for Chiari I-associated syringomyelia over the reviewed time period. Mean±SD age at time of surgery was 11±5 years, eight (38%) patients were male. Syringomyelia was symptomatic in 15 (71.4%) cases. Symptoms attributed to syringomyelia included weakness in six patients (28.5%), paresthesia/anesthesia in 11 cases (52%), dysesthesia in six cases (28.5%), incontinence in four patients (19%), and gait instability in four cases (19%). Six patients (28.5%) had brainstem or cranial nerve symptoms. Tonsil herniation was found more than 5 mm below the foramen magnum and C1 in four patients (19%), and between C1 and C2 in nine (82%) patients. Syringomyelia involved the cervical spinal cord in 17 cases (81%), and the thoracic spinal cord in seven cases (33.3%). Cerebrospinal fluid flow imaging showing slow flow movement of the foramen magnum ventral section, with obvious abaissement of dorsal section, indicated obstruction (Figures 1–4). All the patients showed slow flow movement of the foramen magnum ventral section. Aseptic meningitis was seen in four (19%) patients, wound breakdown was not seen in any patients. However, these perioperative complications did not occur in any patients who experienced syrinx-specific symptom persistence. Furthermore, none of these perioperative



**Figures 1 and 2.** Preoperative MRI shows downward herniation of the caudal cerebellum or medulla oblongata into the spinal canal and Chiari-associated syringomyelia

complications required reoperation for treatment. No patients showed evidence of craniovertebral bony abnormalities, including cranial settling, platybasia, Klippel-Feil, or basilar invagination.

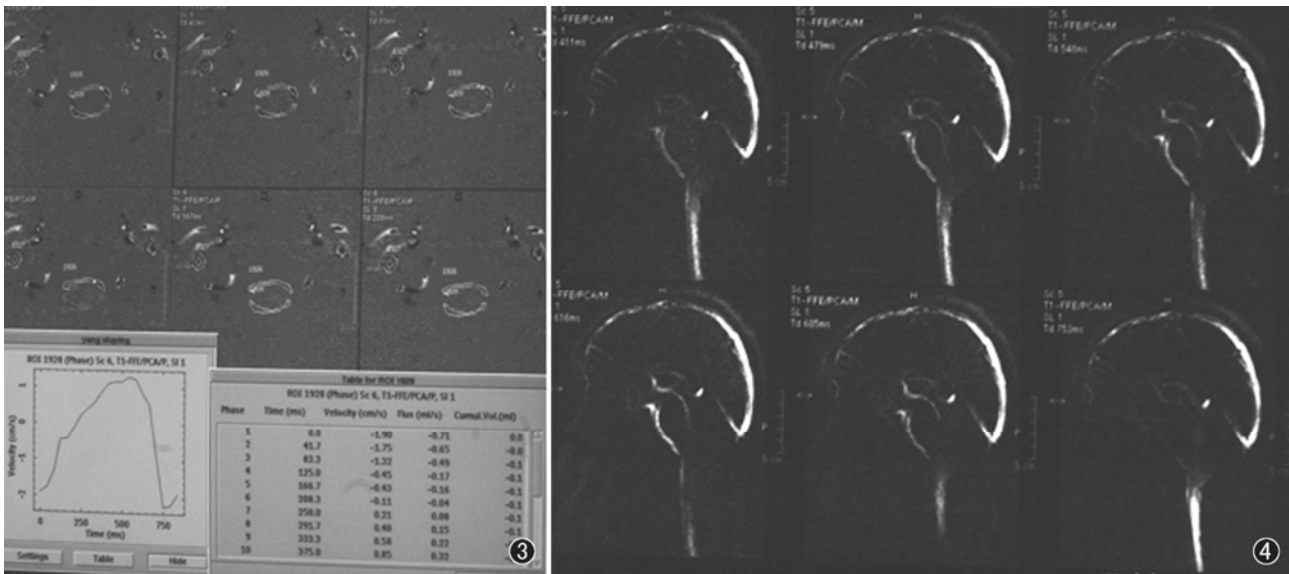
### Response of neural endoscopic posterior fossa decompression and atlas laminectomy

Eleven patients (52.4%) experienced radiographic improvement in syringomyelia (decreased size or resolution) during the follow-up period. Nine patients (42.8%) demonstrated decreased syrinx size and four (19%) demonstrated resolved syrinx. Of the 15 patients with symptomatic syringomyelia, 11 (73.3%) experienced symptomatic improvement. The median time to symptom improvement was four months after surgery. Post surgical MRI examinations indicated complete and sufficient decompression of foramen magnum region (Figures 5–8).

## DISCUSSION

There are different procedures followed at individual hospitals, especially in regards to whether or not the dura mater or arachnoid membrane is cut open and whether or not to perform resection of the amygdala cerebelli.<sup>4-7</sup> Traditional surgery causes maximum surgical trauma, with increased injury to posterior muscles and ligaments. If the patient presents with congenital developmental deformity or instability of the cervical-cranial junction, surgery will affect stability, along with prolonged recovery time and with brace support for up to 3 months post surgery. We carefully analyzed the specific anatomy of this area, concluding that the main predisposing factors were foramen magnum bone structure stenosis or stenosis and obstruction caused by amygdala cerebelli herniation. The main objective of traditional craniotomy surgery is to release compression, increase space and regain CSF flow circulation.

Professor Chiari first summarized over 40 cadaver cases from 1891 to 1896 with amygdala cerebelli herniation verifying metencephal malformation, finally classifying Chiari malformation into four main types. Type I is the most commonly seen, where the amygdala cerebelli



**Figures 3 and 4.** Preoperative cerebrospinal fluid flow imaging shows CSF flow ventral to the cord, cervicomedullary junction and brainstem, but no dorsal flow is seen.

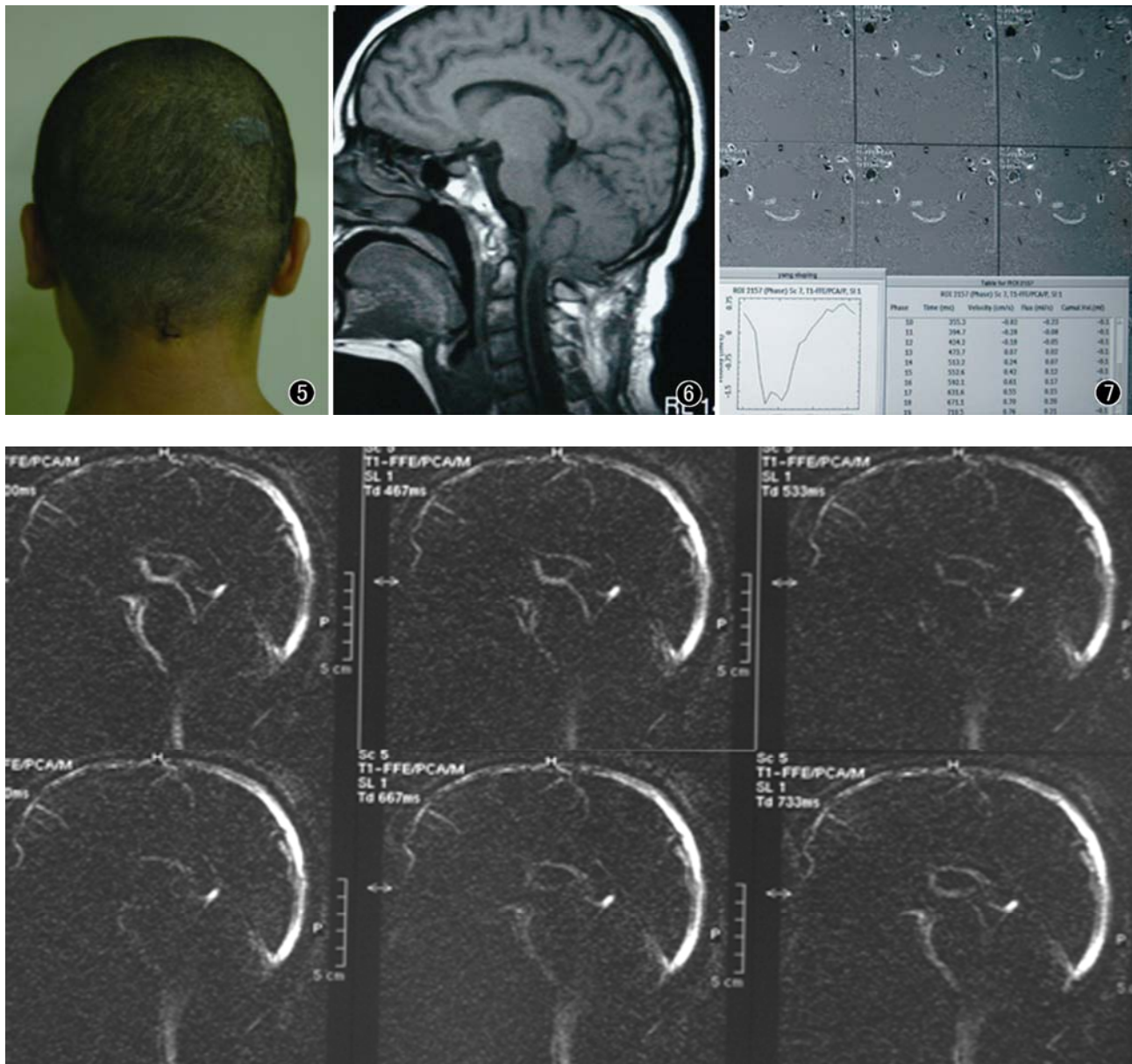
herniation sags deep below the foramen magnum. Even if herniation is up to 7 mm, its contents may still be within the normal range. Today, pathology diagnosis requires herniation of greater than 5 mm. Cases with Chiari malformation type I are most likely to be complicated with decreased posterior cranial fossa volume, mild elongation of the fourth cerebrum ventricle and brain stem, as well as syringomyelia. To further classify type I, Iskandar<sup>8,9</sup> proposed the concept of types 1.5 and 0. Type 1.5 is based on the basics of type I complicated by obvious elongation of the brain stem and fourth cerebrum ventricle. Type 0 has evident syringomyelia, however, without the presence of amygdala cerebelli herniation. but it is necessary to differentiate the above malformations from various tumors and pathogenesis prior to surgery. With such considerations, our particular patient was diagnosed with Chiari malformation type 0.

The reason for the herniation of the amygdala cerebelli remains unknown and there are various hypotheses offered. One that is most accepted is dysplasia of the fetal mesoderm occipital bone, causing constriction of the posterior cranial fossa, thus inducing herniation of the amygdala cerebelli. During infancy, rapid growth and development of the occipital bone somewhat relieves posterior cranial fossa constriction, often referred to as the adolescent stage.<sup>1</sup> Marin-Padilla<sup>10</sup> gave an overdose of Vitamin A to pregnant mice to induce fetal mesoderm dysplasia, provoking herniation of amygdala cerebelli, thereby successfully supporting this particular hypothesis. This, however, only explains a portion of the type I patients, with no viable reasoning or evidence for other types.

A more accepted theory regarding formation of syringomyelia is the “water hammer effect”: Gardner,<sup>11</sup> through extensive research regarding CSF water dynamics, proposed CSF fluctuation induces centralized

expansion and gradual development. Williams,<sup>12</sup> based upon Gardner fundamentals, showed existence of pressure differences between cranial and neural canal CSF. Patients with amygdala cerebelli herniation suffer from CSF circulation obstruction. Quencer<sup>13</sup> used MRI examination to show that patients having undergone decompression surgery had recovery of CSF circulation with obvious shrinkage or disappearance of syringomyelia, hence also supporting the pressure gradient theory.

Atlanto-occipital decompression is generally regarded as an effective surgical treatment of amygdala cerebelli herniation and syringomyelia.<sup>14</sup> As for whether or not the dura mater must be opened, or to cut open the arachnoid membrane, or the necessity of exploration of the fourth cerebrum ventricle or epicele exit are all points of current discussion.<sup>4-7</sup> Likewise, the necessity of excising the amygdala cerebelli and whether or not it is necessary to perform syringomyelia shunting are also factors that are still currently being debated with a variety of supporting arguments. The main pathogenic cause of this disease is absolute bony structure stenosis of the foramen magnum or relative obstruction and stenosis caused by amygdala cerebelli herniation. Many people consider the presence of bone structure induced stenosis to be a requirement.<sup>15</sup> There is an ongoing argument as to whether the dura mater should be opened during surgery.<sup>4,5,7</sup> However, most practitioners disagree and would advise against performing amygdala cerebelli excision and shunting of syringomyelia. Our main objective in using endoscopic surgery is examination of CSF flow to determine smoothness and flow rate through ultrasonography. If CSF flow is smooth then it is unnecessary to open the dura. However, if obstruction or stenosis is observed inducing stagnation or there is disturbed flow, then a Y-shaped incision may be made on the dura mater to allow decompression. Upon re-examination, if flow rate



**Figure 5.** Skin incision is as small as 2.5 cm in length.

**Figure 6.** Postoperative MRI examinations indicates complete and sufficient decompression of foramen magnum region.

**Figures 7 and 8.** Postoperative cerebrospinal fluid flow imaging studies show CSF flow ventral to the cord, cervicomedullary junction, and brainstem and dorsal to the cord with extension around the tonsils.

is still affected then it is advisable to cut open the dura mater for complete decompression, while simultaneously guaranteeing maintenance of the arachnoid membrane. After the dura is cut open, the dura membrane may be left open through a very small incision without requirement of suturing. With experience of over 80 cases, there was no occurrence of CSF leakage or cerebella descensus complications. It shows no difference in clinical or radiological improvement in regards to intra-operative ultrasonographic examination to determine whether or not the dura mater is to be cut open for decompression.

All patients underwent preoperative MRI and cerebrospinal fluid flow imaging as well as postoperative 3-month MRI and cerebrospinal fluid flow imaging to

identify the response of neural endoscopic assisted micro-invasive management of Chiari I malformation. Cine phasecontrast MRI has been suggested being a noninvasive and useful tool for surgical risk stratification and identifying patients that may be optimal surgical candidates,<sup>16</sup> may play an important role in the outcome of decompression surgery related to improving symptoms and restoring normal neurological hydrodynamics in patients with Chiari I malformations.<sup>17</sup> We conclude that neural endoscopic assisted micro-invasive management of Chiari I malformation is effective proving by pre-operative and post-operative cerebrospinal fluid flow imaging.

Endoscopic atlanto-occipital decompression fully utilizes

good lighting offered by the endoscope while performing and investigating deeper regions, clear visualization, and angled scopes that aid in enlarging the visual surgical field. Other obvious advantages include reduced trauma and injury compared to traditional craniotomies, with patients discharged 2 days–3 days after operation. Since there is minimal injury to posterior muscles and ligaments, the patient is not required to wear a cervical brace post surgery.

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