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OPINION & COMMENT

The significance of occipitocervical dura angulation in selection of surgery procedures for Chiari malformation type I

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ABSTRACT

At present, the common surgical procedures for the Chiari malformation type I are comprised of posterior fossa decompression, duraplasty and tonsillectomy. Some neurosurgeons prefer these so called minimally invasive surgeries. However, there are still some failures for patients undergoing the above surgeries in clinical practice. Analyzing causes of many surgical failures, the author put forward the anatomical concept of occipitocervical dura angulation (ODA). The ODA is defined as the included angle between the cerebral dura mater and spinal dura mater at the posterior foramen magnum on the median sagittal plane. For Chiari malformation type I without atlantoaxial instability, the selection of appropriate surgeries and accurate evaluation on the effect of the decompression can be realized after the comprehensive analysis both on the severity of tonsil herniation and the ODA. Tonsillectomy may be needed to add to posterior fossa decompression (PFD) and duraplasty for Chiari malformation type I with the ODA being the larger obtuse angle and/or the tonsil herniation to the level of arcus posterior atlantis.

Previous pathological studies about Chiari malformation have been mainly focusing on tonsilla cerebelli and the ventral structure of the foramen magnum region, such as clivus, atlas and odontoid process complexes [1, 2]. Meanwhile, the characteristics of dorsal structure are typically ignored. There has been a lack of theoretical guidance in selection of surgery procedures for Chiari malformation type I. Research has found that the severity of tonsil herniation was a factor affecting the operative results, whereas it was ill-considered for the selection of surgeries only based on its severity. Therefore, the anatomical concept of occipitocervical dura angulation (ODA) is put forward by the author.

1 Pathological basis and surgical failure causes

The pathology for the Chiari malformation type I is that the malformation of the foramen magnum region, and tonsil herniation bring about congestion of the foramen magnum and the obstruction of cerebrospinal fluid circulation; potential secondary results include syringomyelia and hydrocephalus. The therapeutic principle

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for the disease is decompression, including epidural and intradural decompressions and dural expansive repair, making the cerebrospinal fluid circulation at the foramen magnum unobstructed [3]. At present, the common surgical procedures for the Chiari malformation type I are comprised of posterior fossa decompression (PFD), duraplasty and tonsillectomy [4].

In order to reduce surgery-related complications, PFD plus duraplasty without tonsillectomy has been performed [5, 6]. It has been also reported that the PFD is enough with no need of management of dura maters [7–9]. Although satisfactory effects had been obtained for separately reported surgeries, some results were contradictory [8–10], and there were still some failures for patients undergoing the above surgeries in clinical practice [12–14]. In those cases, what were the causes for surgery failures? And which surgery procedures should be chosen?

Through analyzing the causes of many surgical failures, the authors think that it may be due to the lack of understanding in pathological morphology and pathogenesis of Chiari malformation. It has been considered that the pathological characteristics for Chiari malformation were that, as compared with brain tissues of contents, it possesses a smaller volume of the posterior fossa and the herniation is formed because of the inability to contain the cerebella tonsil. The corresponding therapeutic principle is the decompression to increase the volumetric ratio between posterior fossa and its brain tissues of contents. In fact, however, pathological changes of Chiari malformation including the herniation and malformation of cerebellar tonsil, or congestion of foramen magnum, might not be attributable to the small volume of posterior fossa. Some patients with larger volume of posterior fossa and even with cerebellar atrophy also have tonsillar herniation (Fig. 1, Fig. 2). Most of the herniated tonsil tissues present with degeneration,

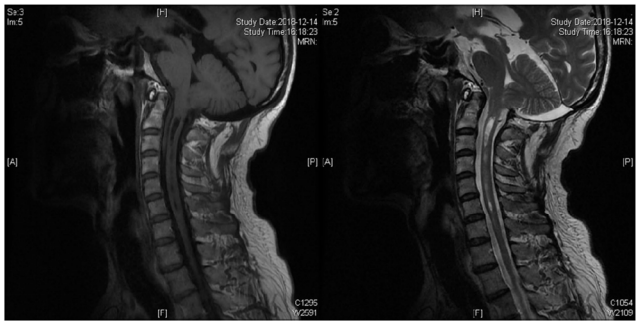

Fig. 1 The MRI of Chiari malformation type I shows that the volume of posterior cranial fossa is significantly larger than that of hindbrain, and ODA was an acute angle.

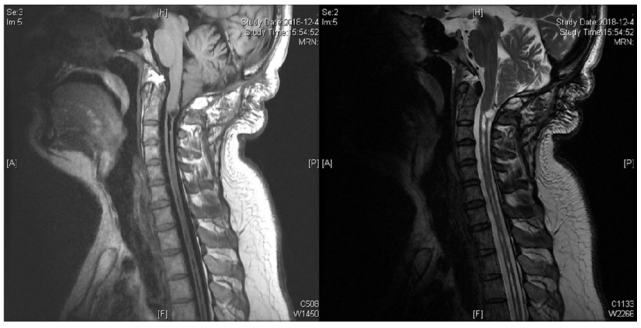

Fig. 2 The MRI of Chiari malformation type I showed cerebellar atrophy and larger cranial fossa space.

scleroses, and adhesion to peripheral tissues, and it is hard to retract even if expanding the volume of posterior fossa (Fig. 3). The key for pathological changes of Chiari malformation is around the foramen magnum. Therefore, the aim of the operation for Chiari malformation type I is to expand the relative volume of the foramen magnum region rather than the whole posterior fossa, making the opening of the fourth ventricle in the reconstructed cisterna magna. But in some surgeries, decompression cannot meet the above goal, and it remains obstructed for the cerebrospinal fluid (CSF) channel, thus leading to surgical failure. Excessive decompression of the posterior fossa may lead to cerebellar ptosis and exacerbation (Fig. 4 A and 4 B) [1]. The selection of appropriate surgical procedures depends on deliberating characteristics of the foramen magnum region of the individual Chiari malformation, especially of ODA.

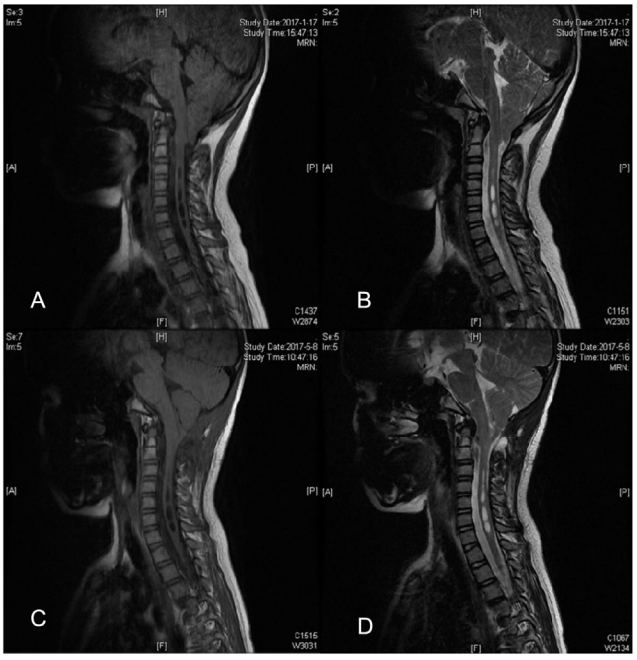

Fig. 3 The MRIs of Chiari malformation type I. (A) and (B) Tonsillar hernia was lower to C1 and occipitocervical dura angulation was a larger obtuse angle before surgery. (C) and (D) Syringomyelia and tonsillar hernia were not significantly improved half a year after PFD plus duraplasty.

2 ODA and selection of procedures

The ODA is defined as the included angle between the cerebral dura mater and spinal dura mater at the posterior foramen magnum of the craniocervical junction on the median sagittal plane (Fig. 5). Normally, the ODA is the smaller obtuse angle. However, when the basilar invagination or platybasia exists, the vertex of ODA moves up, and the angle will be less than or equal to 90°. When performing the proposed expansive duraplasty, the upper end of the preset dura patch is adjusted to be located 2 mm above the cerebellar hemisphere-tonsil incisure of the dura mater, and its lower end is at the level of the arcus posterior atlantis [1]. After the duraplasty, the vertex of ODA would move down to the level of arcus posterior atlantis, and the newly formed ODA would show a significant increase. The triangle formed by both sides of the

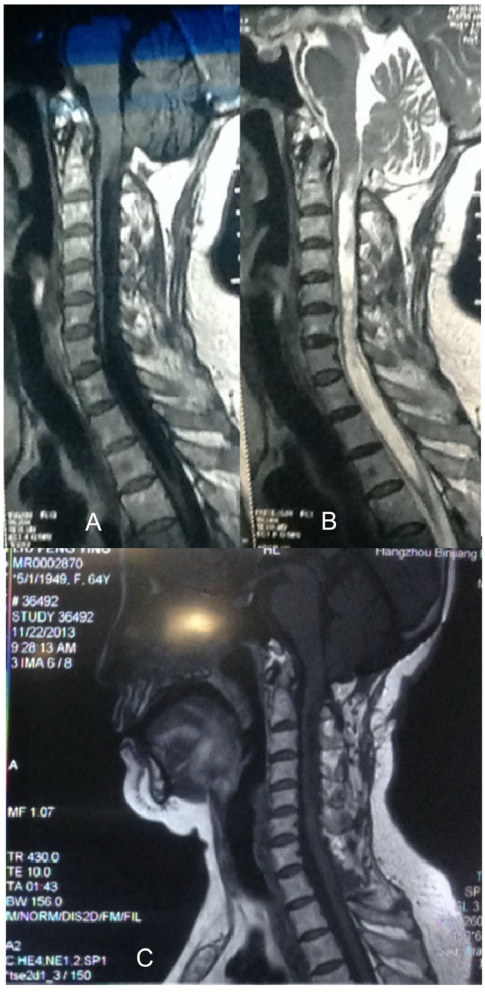

Fig. 4 MRI of Chiari malformation type I. (A) and (B) Cerebellar atrophy and cerebellar ptosis after improper PFD. The ODA was an acute angle and the outlet of the fourth ventricle located below vertex of ODA. (C) After occipital bone repair, atlas posterior arch excision and duraplasty, ODA was increased, hernia and ptosis were recoiled, and syringomyelia became obviously smaller.

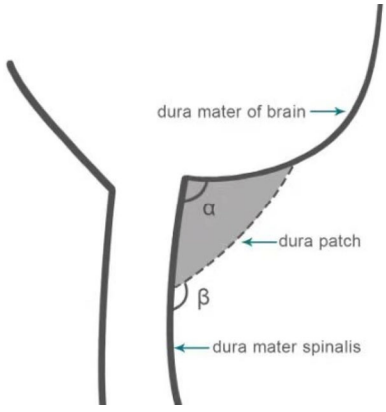

Fig. 5 ODAs before and after duraplasty. Angle α: ODA before duraplasty; Angle β: ODA after duraplasty; Shadow: newly added subdura space by foramen after operation.

original ODA and the dura patch is the newly increased subdural space after the duraplasty (Fig. 5). Obviously, the operation will achieve an increase of intrathecal capacity of the foramen magnum area and reconstruction of the cisterna magna. Therefore, when the ODA is less than or equal to 90° (Fig. 4, Fig. 6), it will have the effect of the newly increased area of the foramen magnum after the PFD and duraplasty; however when the ODA is the larger obtuse angle and the posterior fossa is infundibular, the space of the foramen magnum shows no significant increase even after PFD and duraplasty, and it is hard to retract for the herniated cerebellar tonsil (Fig. 3, Fig. 7). In such case, with relatively severe tonsillar herniation and larger ODA, the PFD with tonsillectomy is recommended, thereby elevating the position of the opening of the fourth ventricle and making it in the reconstructed cisterna magna (i.e., the newly increased subdural area after the tonsillectomy, Fig. 8).

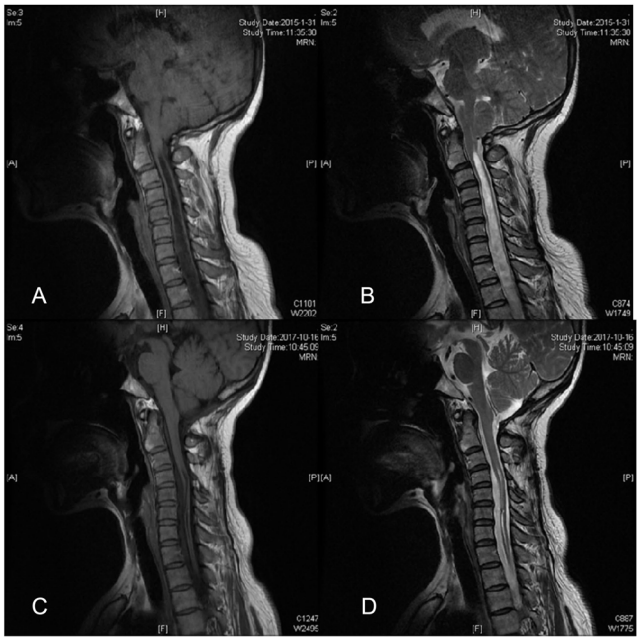

Fig. 6 MRI of Chiari malformation type I. (A) and (B) ODA is close to a right angle. (C) and (D) After PFD and duraplasty, ODA was increased, reconstruction of the occipital cistern was obtained and syringomyelia became obviously smaller.

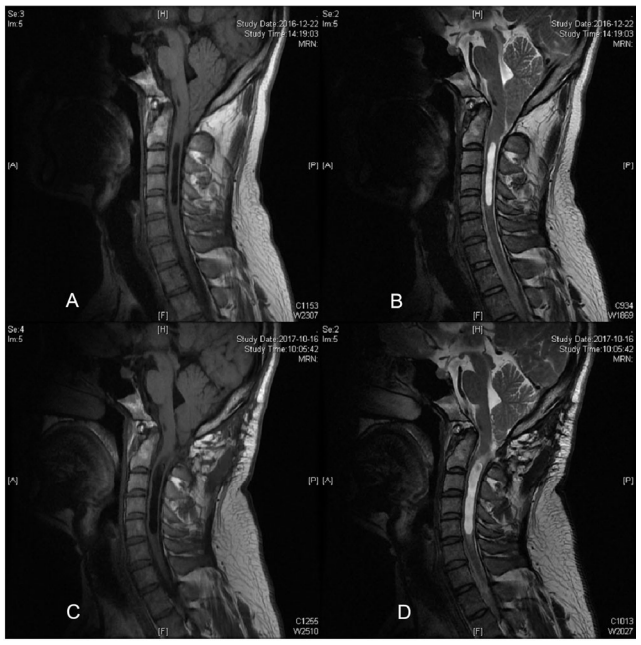

Fig. 7 The MRI of Chiari malformation type I. (A) and (B) ODA was nearly a straight line and its vertex located at the rear edge of the foramen magnum. Posterior fossa was funnel-shaped, with obvious tonsillar herniation. (C) and (D) After PFD with the small bone window plus duraplasty, ODA was about the same as before the operation. The space of the foramen magnum region did not increase significantly, with the tonsillar hernia retracted slightly and syringomyelia not improved.

As seen above, for patients with Chiari malformation type I accompanied by basilar invagination or platybasia, their ODAs were smaller and surgical procedures of PFD plus duraplasty were effective. Meanwhile, tonsillectomy is needed for patients with infundibular skull base and considerably larger ODAs and those with remarkable herniation. Common mistakes should be pointed out that some patients' ODA were smaller, but PFDs were performed only to release the structures above ODA vertex as full as possible, leaving the bone and dura below vertex intact and the remaining outlet of the fourth ventricle located below the vertex. Secondary results were CSF obstruction not being released and symptoms worsen. Revision surgery may be needed for the above-mentioned operations which caused excessive decompression

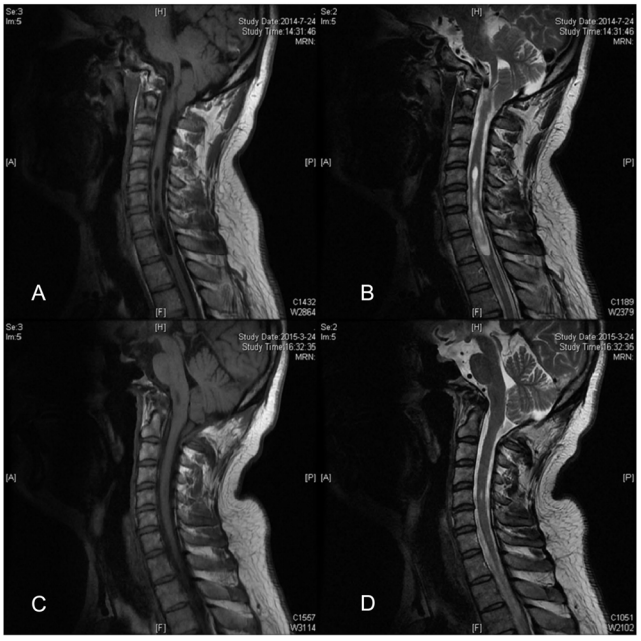

Fig. 8 The MRI of Chiari malformation type I. (A) and (B) Before the operation, ODA was at a large obtuse angle, and the tonsillar hernia was lower to the arcus posterior atlantis. (C) and (D) One year after PFD, duraplasty and tonsillectomy, reconstruction of the occipital cistern was obtained and syringomyelia almost disappeared.

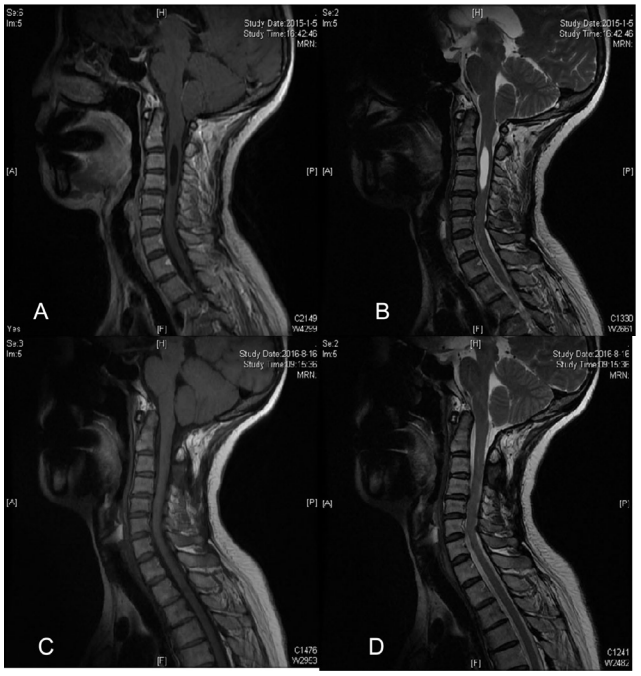

Fig. 9 The MRI of Chiari malformation type I. (A) and (B) Before operation, ODA was smaller, and occipital cistern magna disappeared and syringomyelia was obvious. (C) and (D) One year after PFD, duraplasty and syrinx-shunting, reconstruction of occipital cistern was obtained and syringomyelia disappeared.

and cerebellar ptosis (Fig. 4). Other mistakes include that some doctors overemphasize the decompression of the small bone window for patients with Chiari malformation type I, in spite of ODA dimension and tonsil herniation severity. As a result, the decompression was insufficient and the effect was poor (Fig. 7) [12, 13]. For patients with the tonsil herniation to the level of arcus posterior atlantis or lower, even if the ODA was less than 90°, the obstruction of CSF pathway may still be retained after the duraplasty due to the lower position of the opening of the fourth ventricle. Therefore, tonsillectomy is needed for such cases [13]. Syrinx-shunting may be done in addition to decompression surgery for larger tension syringomyelia (Fig. 9). However, there is no need for immediate treatment for children with Chiari malformation type I without syringomyelia and hydrocephalus; routine observation and regular follow-up are recommended [15].

3 Conclusions

For Chiari malformation type I without atlantoaxial instability, the selection of appropriate surgeries and accurate evaluation on the effect of the decompression can be realized after the comprehensive analysis both on the severity of tonsil herniation and the ODA. For Chiari malformation type I with the ODA being the larger obtuse angle and/or the tonsil herniation to the level of arcus posterior atlantis, tonsillectomy may be needed in addition to PFD and duraplasty.

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Conflict of interests

All contributing authors have no conflict of interests.

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