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Chiari 1 malformation management: the Red Cross War Memorial Hospital approach

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Abstract

Purpose In this paper, we aimed to review our institutional opinions and experience with Chiari 1 malformation management to determine physician practice and outcomes.

Methods Discussion between 3 clinicians about practice preferences and the management of Chiari 1 worldwide. Retrospective review of clinical cases over a 10-year period (2009–2018).

Results Although there are some minor differences between clinicians in our practice, our approach is broadly similar. We treat incidental Chiari 1 malformations conservatively, with clinical and radiological surveillance, reserving intervention for patients who develop clinical signs or radiological deterioration. We prefer surgical intervention for patients with typical symptoms or a Chiari 1 malformation with radiological progression. If symptoms are atypical, we prefer surveillance. Our preferred operation is a conservative suboccipital craniectomy with expansion duraplasty and adhesiolysis. Our operative complication rate was low and there was no mortality or major morbidity in our series. Surveillance for incidentally discovered Chiari 1 malformations has been a safe practice in our experience.

Conclusion Clinical practice among three clinicians in our institution is broadly consistent. We have a conservative approach to Chiari 1 malformation management and our approach appears to have a low morbidity.

Keywords Chiari 1 malformations · Children · Management

Introduction and purpose

Chiari malformation type 1 occurs in about one in 1000 births, with a slight female preponderance [1]. It is a relatively common condition in paediatric neurosurgical practice, with a wide variety of management approaches described. There are many theories about the pathogenesis and pathophysiology of Chiari 1 malformations and the associated syringomyelia. We believe that the most important aspect of the pathogenesis of the condition is a disruption of normal cerebrospinal flow across the craniocervical junction. For the purposes of this paper, we aimed to review our institutional experience with Chiari 1 malformation management to determine local physician practice and outcomes.

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Methods

We discussed the following subjects between 3 clinicians at our institution (Red Cross Children's Hospital, Cape Town): views on Chiari practice worldwide, interpretation of the existing literature, and personal practice preferences. These views are summarised below. We attempted to develop this into an institutional approach to managing patients with Chiari 1 malformations. To examine this against our actual practice, we retrospectively reviewed our clinical cases of Chiari 1 malformation over a 10-year period (2009–2018).

Results

The following summarises our views.

What symptoms are considered typical? Criteria for intervention

We are aware of the many theories on Chiari 1 development, how these have evolved over the years, and how these have

influenced worldwide management. Many neurosurgeons have strong views on who to treat and how to treat, with therapeutic strategies ranging from shunting all patients to doing craniocervical junction fusions in all patients [2, 3]. However, although there are many extreme views, there are many areas where there is a relatively strong agreement. Most neurosurgeons will agree that the presence of a Chiari 1 malformation and a syrinx in close proximity, or a Chiari malformation with classic symptomatology, is a relatively strong indication for surgical decompression. Symptoms that can be ascribed to Chiari 1 malformations are often contentious though. We subscribe to the classical description of symptomatic headache as above [4]. In smaller children and those who cannot verbalise though, we look for signs of meningeal irritation on positional changes, and arching posturing of the neck that is associated with irritability and crying [5]. We consider classical clinical features to include those that reflect brainstem compression (in up to 20% of cases) such as dysarthria, vocal cord palsy, swallowing difficulties, dysphonia, sleep apnoea (central type) [6], and tongue atrophy [5], as well as long tract deficits such as weakness, sensory changes, and upper motor neuron signs [5]. Some researchers have also reported downbeat nystagmus as well as cerebellar signs, such as ataxia, often disappearing after Chiari decompression [7–9]. Syringomyelia may add other clinical features such as a suspended sensory level, atypical patchy sensory changes, or dysesthesia in the limbs and intrinsic hand muscle atrophy [5]. In our experience, this can present atypically, including recurrent ankle sprains in one of our patients and complex regional pain syndrome in another, both of which responded to surgery. Scoliosis is also a finding in up to 30% of patients with syringomyelia [10, 11]. Sometimes, syringomyelia presents with subtle signs. Absence of superficial abdominal reflexes has been described as an early sign of syringomyelia in children with scoliosis [12]. Great clinical acumen and a low threshold for MRI scanning of the whole neuraxis are therefore important.

As is probably true at most institutions, we have increasingly encountered incidentally detected Chiari malformations in recent years. For example, modern head CT scan protocols for head injury (rapid sequence, lower radiation dose, surveillance for retroclival haematomas, and greater awareness) have increased the detection of incidental Chiari malformations after minor knocks to the head. Increasing medicolegal concerns have likely also contributed to a lower threshold for obtaining imaging for relatively minor symptoms that may have nothing to do with the Chiari malformation. Typically, a Chiari malformation is diagnosed by the radiologist when there is cerebellar tonsillar descent more than 5 mm below McRae's line. We pay less attention to the actual measurements and rather focus on the impression of compression at the craniocervical junction and association with symptoms. Occasionally, asymptomatic patients may also have an incidental syrinx, which complicates decision-making. Some of these patients may appear asymptomatic but closer enquiry may reveal a history of clumsiness and frequent falls. It should also be considered that the Chiari may have contributed to the injury.

Institutional approach and type of surgery used

The following summarises the key factors that we felt are important in an approach to managing Chiari 1 patients. A thorough history and examination (general and neurological) is critical, specifically considering syndromes known to be associated with Chiari 1. Subtle clues are looked for with respect to clumsiness, unsteadiness, airway patency and respiratory function (especially during sleep), and bulbar function. Patients with incidental or symptomatic Chiari malformations all undergo whole brain and spine MRI. We always look for hydrocephalus or signs of raised intracranial pressure (ICP) as a cause of tonsillar descent and a tethered cord as a cause of syringomyelia. We examine for signs of intracranial hypotension, as in the case of occult CSF leaks. We do not routinely perform CSF flow studies at the craniocervical junction. Reconstructed bony CT scans are performed if there is a suspicion of an abnormal craniocervical junction, especially if there is ventral compression. Any features on imaging of CCJ instability is followed up with dynamic CCJ imaging—flexion and extension X-rays.

Although not routine, we have a low threshold for performing ICP monitoring and sleep studies. Formal sleep studies are useful when there is equipoise, especially in very young patients. If the study shows central apnoea, it strengthens the argument for decompression. In the case of normal sleep studies in otherwise well patients, we are more inclined to continue clinical and radiological surveillance.

Patients with craniosynostosis all have their craniofacial surgeries performed as the primary intervention and radiological surveillance of the Chiari is performed thereafter. Extra caution is advised for patients with complex craniofacial disorders, especially when there is a morphologically abnormal posterior fossa, as there may be abnormal venous drainage of the posterior fossa, which has consequences for the surgical approach and anticipated morbidity if a posterior fossa decompression is considered.

In the absence of any other cause of tonsillar descent or syringomyelia, the history, clinical examination, and MRI findings will determine the need for surgery. The mere presence of cerebellar tonsillar decent of more or equal to 5 mm below McRae's line is not by itself an indication for surgical intervention. The degree of compression, as judged on Sagittal T2 MRI of the craniocervical junction (CCJ), amount of CSF flow around the CCJ, as well as the presence, or absence, of signal change on MRI in the medulla of the cord or brainstem are all more important factors in our view. Only proven instability of the CCJ will be treated by instrumented fusion in our unit.

In general, a narrowed CCJ due to the Chiari malformation and the presence of a syrinx in the cervical or upper thoracic spine are indications for surgical management, although in truly incidental cases, we first watch the child over some time and repeat the scan. Where symptoms are thought to be attributable to the Chiari or the syrinx, or where there is radiological progression (progressive tonsillar descent, brainstem compression, syringomyelia, and/or scoliosis) in an asymptomatic patient, we offer surgery.

As is the case in any unit where there is more than one surgeon, we have had institutional differences in practice between surgeons and over time. However, our typical surgical procedure for Chiari 1 malformations follows these principles: conservative foramen magnum bony decompression (approximately 2 cm) with removal of the C1 posterior arch; removal of epidural fibrous bands, dural opening, adhesiolysis of any arachnoid scar; and expansion of the dural sac with a bovine pericardial graft with a watertight closure. The syrinx is not treated at the same sitting—we aim to observe the response of the syrinx to posterior fossa decompression. In our experience, most of these will either resolve or reduce in size after adequate Chiari decompression and expansion duraplasty if the CSF flow has been restored. Only symptomatic syrinxes that do not resolve or increase in size are treated with insertion of a syringo-subarachnoid shunt. We will, however, try to avoid this as in our experience, as in most units around the globe; these shunts are fraught with troubles of blockage and migration. We do not routinely cauterise cerebellar tonsils, but have done so in the past for tonsils that descend well below C1 and form part of the compression as seen on MRI. We have not performed instrumented fusion of the CCJ for Chiari 1. We have used intraoperative neurophysiology and intraoperative ultrasound routinely in the past, and now use them for selected patients.

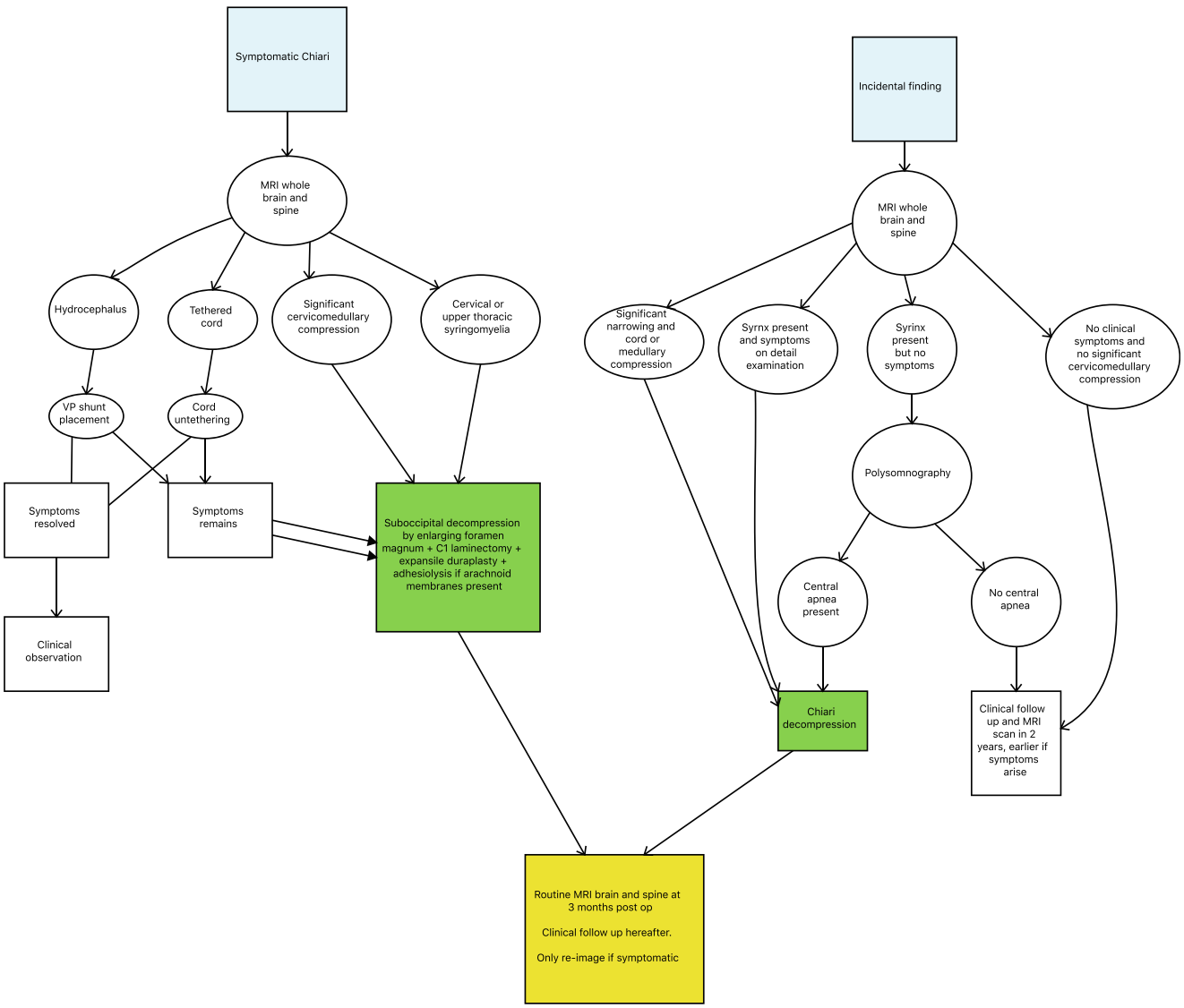

Fig. 1 Therapeutic approach

When is the result considered good?

We focus primarily on the clinical outcome. Our goal is improvement of the presenting symptoms of the patient. In our experience clinical success is more reliable than resolution of radiological characteristics. In our practice, we routinely do a 3-month postoperative brain and spine MRI, which we use as the new "baseline" MRI to evaluate any future symptom-prompted MRI scans. We refrain from "treating the scan" in patients with Chiari malformation and will only intervene if there is continued progression of a syrinx or the patient becomes symptomatic again (or if there has not been symptomatic improvement).

Short presentation of our series

We operated on 25 cases during this period. Our complication rate has been low. We have had no postoperative acute neurological deterioration or new neurological deficits, CSF leaks, wound infections, meningitis, or development of hydrocephalus. We have had no mortality or permanent morbidity. We have had three patients who developed intraoperative complications, one had a significant venous injury, one had haemodynamic instability, and one patient developed significant bleeding from abnormal veins in the neck musculature requiring abandoning of surgery (a patient with a neck lymphangioma). These all resolved without permanent sequelae. Patients are typically managed in the intensive care unit for one postoperative day and we have never needed a longer postoperative stay.

In terms of surgical outcomes, the clinical success rate is high, especially when there are clear symptoms attributable to the Chiari malformation and/or syrinx. Radiological improvement is less than clinical improvement in our experience. Occasionally, tonsillar descent and/or syringomyelia are worse postoperatively. Usually, this stabilises, but occasionally, a second operation is required. In our series, two patients required a second operation—in both of these patients, tight fibrous bands had regrown across the posterior CCJ.

In our institutional experience with incidentally discovered Chiari malformations, several patients (approximately 20%) end up with surgery for the following reasons: subtle clues on history that the patients are not truly asymptomatic, radiological progression, and/or the development of new symptoms or signs. There were no cases who developed acute neurological deterioration during surveillance.

For patients who undergo ICP monitoring (in the absence of hydrocephalus), the vast majority of these show normal ICP.

Our therapeutic strategy is summarised in Fig. 1.

Conclusion

In summary, we have a relatively conservative approach to Chiari 1 malformation management at our institution. We intervene if patients have symptoms clearly attributable to the Chiari/syrinx, or if there is increasing tonsillar descent and/or enlarging syrinx. For incidentally discovered Chiari malformations, we perform clinical and radiological surveillance. Most patients do not require surgery. With this approach, our patient outcomes have been acceptable. We have had no mortality or major morbidity of surgery and we have not had any acute neurological deterioration in patients who we were treating with surveillance.

Compliance with ethical standards

Conflict of interest None of the authors have any conflict of interest to declare pertaining to this article. No funding or special treatment was received in any way whatsoever.

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