A Review of Controversial Topics

Controversies in the diagnosis and management of adult Chiari Malformation (CM; the Chiari I malformation) and syringomyelia abound. They include, but are not limited to, those listed in outline form below. This discussion does not focus on the Chiari II malformation, which is associated with other congenital conditions and malformations.[45,46] A brief discussion of each controversy is provided, with selected references for further study if desired.

1. ***Indications for surgery for CM*** - The etiology (cause) of CM varies (and is often unknown), as do the indications for surgery.[1,8,16,28,38,39,40,44,49,52,67] The presence of a Chiari malformation DOES NOT, in and of itself, provide sufficient rationale for an indication for surgery. Most neurosurgeons reserve surgery for the symptomatic patient with correlative imaging findings.[6,17] Symptoms consistent with brainstem compression (swallowing difficulties, diminished gag reflex, speech difficulties, etc) or an associated symptomatic syringomyelia (signs of spinal cord dysfunction, such as weakness or gait disturbance) are usually requisite findings that suggest an indication for surgery. In addition, signs of an intracranial/spinal pressure differential (tussive [cough related] headaches,[59] the presence of an associated syrinx, etc) are often considered indicators for surgery. Vague symptoms such as chronic fatigue, non-specific headache symptoms, general aches and pains, etc are generally not considered to be associated with a surgically treatable Chiari malformation.[1] Many surgeons utilize cine flow studies (a modification of a routine MRI that assesses CSF flow in the spaces around the CM) as a tool to determine indications for surgery.[3,9,13,20]
2. ***Indications for surgery for syringomyelia*** – The indications for surgery for syringomyelia, likewise, vary widely. In the presence of a Chiari malformation, the

syrinx may be effectively treated by a Chiari decompression procedure.[7,42] In the absence of a Chiari malformation, trauma, prior hemorrhage, prior surgery, or prior meningitis may be implicated.[11,12] Oftentimes, the etiology or cause of an isolated syrinx is unknown. Regardless, management is controversial and depends on several factors including age (for example, less likely to treat asymptomatic adults than asymptomatic children), presence or absence of symptoms, size of the syrinx, whether a readily treatable cause such as CM or a tethered cord is present.

3. ***Indications for occiput to spinal fusion or C1-C2 fusion***, with or without anterior decompression (usually via the transoral route) – Occiput to spinal or C1-2 fusion for Chiari malformation is rarely indicated. Such may be the case when severe basilar invagination (angulation of the skull base with respect to the axis of the spine), in which anterior brainstem compression is present at the base of the skull (foramen magnum).[18,26] Such a fusion is clearly indicated if an anterior decompression via the transoral resection of the odontoid process (top portion of the C2 vertebra) is performed.[7] Controversies arise depending on the extent of skull base angulation (e.g., how severe should the angulation be before a fusion is recommended?), symptomatic state, response to prior surgery for Chiari decompression, and others.
4. ***Indications for tethered cord release*** – A tethered spinal cord is uncommonly associated with CM or syringomyelia. When a tethered cord is obviously present on MRI, surgery is often indicated to prevent neurological and bladder dysfunction. Recent claims that tethering of the spinal cord may occur in CM patients without MRI evidence of "occult tethered cord" are fraught with controversy.[41,56]

5. Operative strategy options for CM

a. *The need for bony decompression* – When a Chiari decompression is indicated, bony decompression of the foramen magnum and C1 arch is thought by most surgeons to be necessary. This opinion, however, is not universally held.[64,65]

b. *The need for dural opening* - After bony decompression, opening the dural covering of the foramen magnum is thought to be necessary by 80-90% of neurosurgeons. The rest, however, are convinced that bone decompression is all that is required.

c. *Arachnoid opening* – After opening of the dura mater during a Chiari malformation operation, many surgeons attempt to preserve the integrity of the arachnoid in order to minimize surgical manipulation and subsequent scarring in the subarachnoid space. Others feel that such is necessary in order to adequately decompress.

d. *Tonsil coagulation* – Of the surgeons who recommend arachnoid opening, many recommend tonsil shrinkage by coagulation or other means. The tonsils represent the lower portion of the cerebellum that descend through the foramen magnum in a Chiari patient. Those who argue for such a procedure state that decompression of the 'crowded space' in the region of the foramen magnum is enhanced by such a technique. Those who argue against this strategy, state that scarring is induced in the subarachnoid space by such a surgical intervention.[19]

e. *The use of traditional open versus endoscopic approaches* – Opinions vary regarding the use of traditional open versus endoscopic approaches to CM. Although most surgeons utilize the open approach, some state that the same

results can be obtained with less invasive approaches. Obviously, an endoscopic approach has limitations regarding bone removal, etc. If such is not felt to be relevant in a specific case, many would consider a less invasive endoscopic approach. The major controversy regarding the endoscopic approach, however, is that it consists of endoscopic tonsillar shrinkage with minimal bone removal and dural opening. Some contend that the long-term possibility of scar formation secondary to tonsillar shrinkage has not yet been adequately addressed to warrant wider use of this procedure.

f. *Dural closure* – When the dura is opened, most surgeons recommend dural closure at the end of the procedure. A minority of surgeons, however, state that such closure is not necessary and may even increase the risk of dural scarring to underlying neural structures. Interestingly, neither technique has been shown to be superior so far.

g. *Indications for the use of a dural patch* – When dural closure is performed, a patch is usually placed, which expands the dural sac, presumably allowing for a more depends usually dependent on surgeon preference.[23]

h. *The choice of dural patch material* – Many patch materials are available, ranging from autogenous (local, patient's own) tissue to a variety of cadaveric, animal, and artificial materials.[10] Local patch material can be obtained from the pericranium (tissue overlying the skull), ligamentum nuchae (midline tissue in the neck), and other sources.[15]

i. *Indications for a simultaneous syrinx shunt* – Most surgeons, when faced with a CM and an associated syrinx, simply decompress the CM. Some, however,

recommend shunting the syrinx as well. Those who simply decompress the CM argue that the shunting procedure adds unnecessary risk.

j. *The extent of bony decompression* – Currently, most surgeons decompress the occiput to a lesser extent than in years gone by. Usually about a 1 x 2 cm portion of occiput, as well as the lamina of C1, are resected. Extensive resection of occipital bone can result in recurrent and relatively refractory symptoms due to the sagging of the cerebellum (cerebellar ptosis).[24]

6. ***Operative strategy options for syringomyelia***[4,29,34,35,47,48,61,69] – The etiology (cause) of syringomyelia is not always obvious. Tumor,[57] trauma,[55,62,63] and CSF inflammatory process are but a few. Most often, however, the diagnosis is not known and the syrinx is stated to be idiopathic in nature. To complicate matters further, syringomyelia has been observed in rare circumstances to spontaneously regress without treatment.[66]

    i. *Syrinx shunting procedures* – Although the mainstay of treatment for idiopathic or posttraumatic syringomyelia for years, shunting procedures have been shown in recent years to be fraught with peril and have therefore fallen into disfavor.[55,60,70] Most surgeons shy away from such strategies due to low long term efficacy, risk of spinal cord injury during placement and the inability to detect shunt patency postoperatively. Nevertheless, shunting procedures are appropriately employed in selected cases. Several types of shunts may be used. These are detailed in another section and have their own inherent controversies: Syringo-subarachnoid shunt – Syringo-subarachnoid shunts drain the syrinx fluid into the spinal

fluid space (subarachnoid space). This procedure is the easiest to perform of the three.[2,14,22,30,51]

ii. Syringo-pleural shunt – Syringo-pleural shunts drain the syrinx into the space outside the lung. Pneumothorax and hydrothorax are complications of this procedure.

iii. Syringo-peritoneal shunt – Syringo-peritoneal shunts drain the syrinx fluid into the peritoneal or abdominal cavity.[33,54]

b. *Procedures that restore CSF pathway patency* – Trauma, meningitis, previous surgery and others may cause scarring in the CSF pathways (arachnoiditis). In *turn, this arachnoiditis* may cause blockage of CSF flow and resultant neurological dysfunction with or without syringomyelia. The restoration of CSF flow by creating a larger space (via a laminectomy or a CM decompression and the placement of a dural patch graft)[53,68] may be helpful, even in cases where there is no obvious CM.[27] The maintenance of restored CSF flow, however, is often problematic.[21]

c. Adjuncts to subarachnoid CSF flow re-establishment strategies

i. The use of scar inhibition strategies – Scar inhibition strategies may be employed following a CSF restoration procedure. Such techniques include the application of scar inhibiting material such as collagen-based dural graft matrices.

ii. The use of a dural patch – A dural patch is usually employed with CSF flow restoration techniques. Such provides more room for CSF to flow.

iii. The management of arachnoiditis, syringomyelia and recurrent syringomyelia – The management of recurrent syringomyelia in the face of arachnoiditis is fraught with peril and is therefore very controversial. The employment of the aforementioned techniques may be useful, but the outcome is often suboptimal.[31,32,36] Surgery, therefore, is usually reserved for the neurologically deteriorating patient.

d. *Spinal decompression and deformity correction strategies* – Sometimes, syringomyelia is associated with spinal column deformities (scoliosis, kyphosis, etc.). It is often difficult to know whether the deformity is the cause or result of the syrinx. It may also be difficult to sort out whether the neurological dysfunction caused or was the result of the spinal deformity. In such complex cases, spinal decompression and deformity correction strategies may be indicated as adjuncts to syrinx treatment.[5,37,43,50,58]

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