

Definitions

achondroplasia - genetic birth defect involving abnormal bone growth

cerebellar tonsils - portion of the cerebellum located at the bottom, so named because of their shape

cerebellum - part of the brain located at the bottom of the skull, near the opening to the spinal area; important for muscle control, movement, and balance

cerebrospinal fluid (CSF) - clear liquid in the brain and spinal cord, acts as a shock absorber

Chiari malformation - condition where the cerebellar tonsils are displaced out of the skull area into the spinal area, causing compression of brain tissue and disruption of CSF flow

contused - bruised

cine MRI - type of MRI which can show CSF flow

decompression surgery - common term for any of several variations of a surgical procedure to alleviate a Chiari malformation

dura - thick outer layer covering the brain and spinal cord

duraplasty - surgical procedure where a patch - or graft - is sewn into the dura

ectopia - abnormal position; refers to the cerebellar tonsils being displaced

fascia - fibrous connective tissue

foramen magnum - large opening at the base of the skull, through which the spinal cord passes and joins with the brain

incidental - in this context, a medical finding that may not be important and was not being looked for

intracranial hypertension - condition where a person's intracranial pressure is

On March 16th, Dr. Stephen Fletcher, chief of pediatric neurosurgery at Memorial Hermann Children's Hospital and section chief of pediatric neurosurgery at The University of Texas Medical School at Houston, will perform a Chiari decompression surgery broadcast live on the internet. During this unique event, Dr. Fletcher and his colleagues will also respond to questions submitted via email.

Despite his hectic OR schedule, Dr. Fletcher graciously agreed to share some of his extensive insight and experience in treating Chiari. To learn more about this exciting event, and Chiari in general, we put Dr. Fletcher, In The Spotlight:

How did the idea to webcast a Chiari decompression come about and what do you hope to achieve by doing this?

F: We decided to do a webcast on Chiari as part of an information program for the Memorial-Hermann Hospital system here in Houston. This is one of the larger hospital systems in the country. The Memorial-Hermann Children's Hospital is rapidly growing and part of that growth is the establishment of the Memorial-Hermann Pediatric Neurosciences Center. The University of Texas Medical School at Houston has had a well established pediatric neurology program for decades and pediatric neurosurgery has been solid here for years. The Texas Comprehensive Epilepsy Program under the auspices of James Wheless is probably one of the busiest epilepsy surgery programs in the country. We may boast the largest pediatric head injury center in the country. So as we enter a new phase of development the hospital thought it might be reasonable to present an informative program on a subject that is getting more press even locally.

About how many Chiari patients per year does your group treat?

F: We average about 30 Chiari decompressive surgeries a year excluding those associated with myelodysplasia. I would say our ratio of visit to surgery is about 4 to 1 at present. Most of our cases are seen by pediatric neurology before they get to the neurosurgeons. Ian Butler, M.D. chief of the pediatric neurology section, will see most of these patients.

Realizing that every case is unique, in general, what are your criteria for recommending surgery to treat Chiari?

F: Operative criteria include presence of symptoms, associated pathological exam findings, and imaging that fits the problem. Sometimes we will operate on patients that have severe imaging findings with a paucity of symptoms or exam findings. Post traumatic patients with incidental Chiari and a contused medulla or upper cervical cord is an example.

There are many variations within the general decompression surgery, what technique do you prefer and why? For example, do you always open the dura, do you ever remove the tonsils?

F: Standard suboccipital craniectomy, foramen magnum decompression and laminectomy as needed is the standard and what we prefer. For the most part, I choose to perform a duraplasty but have not in certain cases. I do not remove the tonsils. We use SSEP's extensively since this institution was instrumental in using that modality even back in the 80's and UT published extensively on the value of neurophysiological monitoring for neurosurgery. (Goldie,W, Butler, I, Miner, M). I feel it is a necessary adjunct to surgery. I have had a few instances where the bone decompression resulted in normalization of the SSEP's and therefore I basically did not put in a graft but scored the dura. The patient improved and has remained stable.

If you do perform a duraplasty, what type of graft do you prefer and why?

F: I am old fashioned and used to use fascia but now use artificial dura. Cadaver dura is used some but a new product from Medtronic called Durepair seems to work for me. Since we neurosurgeons are so quirky I prefer this because it holds a stitch well over others I have tried but don't think variances are worthy of mention.

Is there an optimal age for a child to have surgery?

F: A newborn would likely have a delayed surgery just for practical reasons of weight, blood loss, infection, etc. For the most part timing of surgery will depend on the clinical situation.

How do you determine what level of activity a child can return to after surgery? Would you ever allow a patient to play football after surgery?

F: I am opposed to organized sports in which head contact is frequent. You mention football but soccer is just a bad when kids head the ball. I am not popular with parents on this issue.

Are there any differences in performing this operation on a child as opposed to an adult?

F: For the most part, kid Chiari surgery is easier due to the size of the cervical musculature. The technique is pretty much the same. We usually use the microscope for sewing in the dura but again a quirk.

In general, do you think children with Chiari (Type I) have better outcomes than adults?

F: The outcome is probably based on accuracy of diagnosis. As you are aware there is an association of migraine and pseudotumor (elevated intracranial pressure) with Chiari malformation. Not all patients have this

chronically, abnormally high

intracranial pressure (ICP) - the pressure of the CSF in the skull, or cranium

laminectomy - surgical removal of part (the bony arch) of one or more vertebrae

MRI - Magnetic Resonance Imaging; diagnostic device which uses a strong magnetic field to create images of the body's internal parts

pseudotumor cerebri (PTC) - another name for intracranial hypertension

SSEP's (somatosensory evoked potentials) - type of monitoring which uses electricity to stimulate nerves and then measures the response

suboccipital craniectomy - surgical removal of part of the skull, or cranium, in the back of the head, near the base

syringomyelia - neurological condition where a fluid filled cyst forms in the spinal cord

problem fortunately. We tend to be very conservative in recommending this operation in pure headache patients until the neurologists have had a go with conservative care. Every group seems to have their magical cocktail of drugs, observation, etc to see if they can alleviate the headache. But the caveat is that these patients ARE WORKED UP IN A STANDARD MANNER before assuming conservative care. MRI with CSF flow study, SSEP, entire spine survey to look for syrinx, hydromyelia, tethered cord, Flexion/extension SSEP's if needed, flexion/extension mri if needed, plain films of cspine, formal neurological exam, neurodynamic studies if symptomatic. I will not operate on patients without ssep and flow study.

How do you define a successful surgery?

F: Successful outcomes are based on alleviation of symptoms if that was the indication. If a prophylactic operation is done (again the incidental finding with mri evidence of contusion), then success is harder to define. The more parameters you evaluate prior to operation that will be assessed postop can certainly help in defining success.

Why do you think there are still so many controversial issues regarding Chiari surgery?

F: I am not sure there are that many controversial issues in Chiari compared to other medical entities. Variation in operative technique probably is less important than being able to judge who needs surgery. This usually changes with experience. Treating headache alone with operations is a tricky entity. Review the history of occipital neurectomy for headache or the controversies surrounding spine fusions for chronic neck pain and headache.

What do you think the future holds for treating Chiari?

F: There is a thick transverse band at the cervico-medullary junction in the epidural space that is present in all patients. We are looking at that to see if it varies histologically from normals. I think it has to be removed. I send it to pathology in all cases. We may start doing electron microscopy to evaluate it further. I do believe the CSF flow studies are important. I have seen redo patients that have good posterior flow established that never improved clinically. Why is that? We plan to look at intracranial pressure pre and post decompression by monitoring with a fiber optic monitor during the surgery.

We soon plan to obtain CT of the skull base to look at the various foramen of the skull and see what variants may occur there. Is there constriction of venous outflow? We base this on our experience with achondroplasia. Is this a segmentation problem of the spine. Genetics may be of value here. Why do acquired Chiari patients (low lying tonsils) improve spontaneously in patients with lumbo-peritoneal shunts?

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