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Definitions

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

cine MRI - type of MRI which can show CSF flow

compliance - a measure of a container's stiffness; mathematically expressed as the change in unit volume per unit pressure

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

etiology - factors that contribute to the cause of a disease

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

hydrocephalus - condition where there is an abnormal build-up of CSF in the brain

impaction - when two things are pressed closely together, or compressed

Most people with Chiari and/or syringomyelia will see a neurosurgeon at some point in their life. Neurosurgeons are highly educated and trained professionals, so for patients the experience can sometimes be intimidating. And for those who require surgery...they will find themselves having to put a tremendous amount of faith into the hands of someone they have only met a couple of times and barely know.

How do neurosurgeons decide when to recommend surgery? Why is there so much variation in the technique used among surgeons? What should a patient ask their surgeon before agreeing to surgery?

Dr. Ghassan Bejjani is a neurosurgeon and a Clinical Assistant Professor of Neurological Surgery at the University of Pittsburgh. A man of tremendous energy and enthusiasm, Dr. Bejjani has seen numerous Chiari patients and his current research interests include why Chiari surgeries fail and the relationship between Chiari and intracranial hypertension.

We put Dr. Bejjani Into The Spotlight...

Approximately how many Chiari patients do you see a year?
B: Probably every week I see 2-4, so 100-200 per year.

In your estimate, what percent of people with Chiari end up having surgery?
B: One out of three, maybe one out of four. It depends on what you call Chiari; a lot of the borderline cases that may be called Chiari are non-surgical.

What are your general guidelines for deciding if someone should have surgery?
B: I operate only if there are typical symptoms and there is no other etiology for the symptoms, such as coexistent systemic or neurological diseases, secondary gains or psychological issues. On one side, the symptoms of Chiari can be similar to those seen in a variety of systemic, traumatic, neurological as well as psychiatric entities; and on the other "radiologically" significant Chiari malformation may be completely asymptomatic. There really aren't a lot of objective findings you can rely on to confirm your diagnosis, other than the lack of venous pulsations that are not very specific. Also I take into consideration the severity of the symptoms and how they are affecting the quality of life of every individual patient: some can deal with the symptoms without any major handicap, while other are severely disabled by their disease. And the last factor is the morbidity of the surgery.

Do you do a cine MRI for everyone?
B: I do a cine for everybody, but I'm not sure about its true clinical significance at this point. It is however, becoming one of the various elements used in the decision-making process, although the main factor is still the overall clinical impression.

So you form an impression between your neuro exam and the symptoms they report?
B: Yes, especially the symptoms. I look at the symptoms mainly. I take into consideration the spontaneous venous pulsations (SVP), I look at the MRI, and I look at the psychological background. Is the patient psychologically stable? And that's where the main trick is, because symptoms can lead to secondary psychological problems. That's one thing I'd like to research in a surgical outcome study. Do psychological profiles differ before and after surgery, because symptoms can put you on the edge and lead to secondary psychological alterations. I saw a lady today in the office who had typical Chiari symptoms for 15 years a typical Chiari on MRI, and she's been told she's crazy for the past 10 years. Your affect becomes different when everyday, everyone is telling you this. So you have to put everything into context. Then I tell the patient about the potential outcomes of surgery and the potential risk. In my series overall, the risk of significant adverse complications is less than 5%, and our CSF leak rate has gone down to 0% over the past three years. When you see a lot of patients, you get an idea of who will do well with surgery, although there are always surprises. What's needed is a scientific outcomes study to determine who does well with surgery and why. Once there are outcomes studies, we can move to a more scientific approach in dealing with Chiari.

Does the surgical technique you use vary much from patient to patient?
B: In 85-90% of patients I use the same technique. In the remaining 10-15% I might do something different.

What might one of the different things be?
B: Sometimes, I don't open the dura. For example in some cases of Hydrocephalus where the ectopia might be secondary and the risk of a leak is high. Or for a patient who only has symptoms

with extreme activity or neck movement, like a young athlete. But most of the cases are performed with similar techniques.

What type of graft material do you use?
B: The one that I like the most is Durasis. It is made from porcine intestinal submucosa. We just finished a study on it. We had 30 some patients with no CSF leaks and only two resolving fluid collections. It's strong, easy to handle, very easy to suture without having to worry about suture hole elongation and is not rejected. It will be on the market soon.

Why do you think there is so much variation in the surgical technique used among surgeons?
B: Treating Chiari is largely an art, it's not completely scientific yet. There are currently no prospective, outcome studies to determine which technique is better.

What percent of Chiari patients become symptom free after surgery?
B: I'd say probably 50%-60% become completely symptom free although another proportion improves significantly. In some subgroups of patients, the odds of success can be much higher or much lower**.**

What percent of syringomyelia patients become symptom free after surgery?
B: I'm actually seeing less syringomyelia with Chiari because the Chiari is being found earlier**.** With the widespread use of MRI, more isolated Chiari are being discovered, whereas in the past, mostly Chiari patients with Syringomyelia had symptoms and signs that were severe enough to justify obtaining the invasive diagnostic studies used back then (encephalography, myelography). The ones I do see have probably been there awhile. It used to be that the majority of patients had SM, now it is shifting to a much smaller percentage. The problem with SM is how long have the symptoms been there. If they've been there a long time, they probably aren't going to go away completely. But stabilization and some improvement of the symptoms will probably occur 70%-85% of the time. The odds of complete symptomatic resolution are probably small, especially in advanced cases.

Many surgeons describe the decompression as technically simple, yet at the same time say that success is highly dependent on individual surgical skill. Any thoughts on this?
B: I disagree with that. I do not think that the individual skill of the surgeon is the most important factor in determining the success rate. I think other factors play a major role, mainly the clinical decision-making. It is there that the experience of the surgeon becomes important: the more patients you see, examine and follow in a thoughtful way, the more you can have an idea about the natural history and outcome, and the more reliable is your estimate of the success rate. It depends on the surgeon's technical skills only if they don't do enough of them, to the point where they have a high complication rate.

Do you think most surgeons are capable of doing a good decompression?
B: Yes. Technically it's not a very demanding procedure. Now, if you go to someone that only does one or two a year, then he doesn't have enough number of patients to have a clear idea of his outcomes and change his techniques accordingly. Another tendency that I have noticed, is that surgeons who do not do a lot of decompressions will do a smaller decompression, people who do a lot of them will do a more aggressive decompression because they are more comfortable. Does it make a difference in the outcome? I don't know. As far as fluid leak, if you do enough of them you learn how to prevent them and hopefully will decrease your leak rate.

How many decompressions do you think a surgeon has to do before they really become comfortable with this kind of technique?
B: It's not that demanding, so maybe 10. The main thing is the clinical judgment. What are the odds of someone getting better and when should surgery be done. Its not life threatening, so at what point do you take the surgical risk?

Do you have any general thoughts on Chiari?
B: I think there are two pathologies involved in Chiari. One is a mismatch between the skull and the brain, so it's a compliance issue. You have a small skull or too much brain leading to a compliance issue. The other pathology is impaction at the foramen magnum, where you have slumping of the brain into the spinal canal, affecting the CSF dynamics. That's why surgery has two goals. One, make the foramen magnum bigger and relive the impaction and flow blockage. Number two make the posterior fossa bigger and correct the mismatch. In my opinion, in some patients, there's more than just compression at the foramen magnum. That's why so many Chiari patients have elevated ICP. Sometimes, when I first slit the dura, I can see the tonsils moving in a dynamic fashion, and when I cut the dura wide [the pressure is reduced] and the movement stops. The cine MRI tells us that the CSF flow in a Chiari patient is very hyperdynamic. And the reason is you have a very stiff system. A non-compliant system is a very stiff system. The system is stiff because there's too much brain or too little skull. That's also why there is overlap between Chiari and pseudotumor cerebri. In PTC the brain is engorged, or swollen. That's why the symptoms overlap.

Do you think compliance is the trigger to go between asymptomatic and symptomatic?

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

meningocele - abnormal collection of CSF surrounded by dura

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

neural hydrodynamics - the study of the flow of fluids - blood and CSF - in the brain and spinal cord system

posterior fossa - depression on the inside of the back of the skull, near the base, where the cerebellum is normally situated

pseudotumor cerebri (PTC) - another name for intracranial hypertension

spontaneous venous pulsations (SVPs) - periodic changes in size of the veins in the retina (in the back of the eye) observable in a large percentage of people

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

ventricle - CSF filled spaces in the brain

Ghassan K Bejjani, M.D.
Clinical Assistant Professor
Department of Neurological
Surgery University of Pittsburgh
Medical Center

Qualifications:

- Authored more than 60 books, chapters, and papers
- Former Visiting Cerebrovascular Fellow, Yale University
- Former Visiting Microsurgery Fellow, Little Rock, AK

Education:

- **St. Joseph University**, M.D.
- **Henry Ford Hospital**, Surgical Internship
- **George Washington University**, Neurological Surgery Residency

Research Interests:

- Adult Chiari malformation
- Syringomyelia
- Intracranial Hypertension

Selected Publications:

- Bejjani GK. **Association of the Adult Chiari Malformation and Idiopathic Intracranial Hypertension: more than a coincidence.** Med Hypotheses. 2003 Jun;60(6):859-63.
- Bejjani GK, Cockerham KP, Rothfus WE, Maroon JC, Maddock M. **Treatment of failed Adult Chiari Malformation decompression with CSF drainage: observations in six patients.** Acta Neurochir (Wien). 2003 Feb;145(2):107-16.
- Bejjani GK, Nora PC, Vera PL, Broemling L, Sekhar LN. **The predictive value of intraoperative somatosensory evoked potential monitoring: review of 244 procedures.** Neurosurgery. 1998 Sep;43(3):491-8.

B: Yes, I think it's a physical problem, it's a compliance issue. Now there is some thought that minor trauma can lead to a venous compromise and subsequent brain engorgement that will make things worse. Women's symptoms get worse before their periods. With water retention, the tissues are swollen, including the brain and the system becomes stiff, therefore triggering the symptoms. There is a whole concept of neural hydrodynamics which will really push things further. We are having a conference in California in May about abnormal neural hydrodynamics.

Given enough time, do you think everyone with a Chiari malformation would develop a syrinx, or is there another mechanism at play that varies among people?
B: No. I think it's a problem with the tissue. The less compliant the system, the more prone you are to develop a syrinx, but I also think tissue can be more prone to develop a syrinx in some people. Maybe in 30 years we'll figure it out.

Do you think stem cells offer much hope for treating the nerve damage caused by a syrinx?
B: I don't think so at this point. It may get there eventually, but I think we're far from that point. I don't see that in the near future.

Do you treat the residual symptoms of your patients - such as neuropathic pain - or do you refer them to another practitioner?
B: I refer my patients to a pain clinic or a specialist.

What questions should a patient ask their surgeon before surgery to ensure they are getting good care?
B: One: what are the odds of success? Two: what are the odds of complications? Three: what are the surgeon's own experiences in regards to complications? You certainly want to know how many they do a year. Make sure they don't just do one every 4-5 years. But that alone is not an absolute rule, because there are very good surgeons that could do a very good decompression, they just don't do a lot of them.

How do you think treating CM/SM will evolve over the next 5-10 years?
B: I think we're about 10-15 years away from really understanding Chiari. I'm hopeful and really optimistic. There are a lot of good young researchers doing excellent work in this arena. That's why bringing these people together at a conference is so important. I think bringing people together that work on different aspects of neural hydrodynamics, be it CSF flow block, elevated ICP, enlarged ventricles, will help expedite an understanding of Chiari.

Do you think there will ever be a non-surgical solution for either Chiari or syringomyelia?
B: I think some group of Chiari patients will eventually be treated medically. These are the ones that are borderline Chiari/possible PTC. In PTC, 6% of patients have Chiari. These are some of the patients that are failing with decompression, and I think these are the patients that will be treated with a drug. For those with a small posterior fossa, I think it will always be a surgical solution.