

Chiari & Syringomyelia News

Delivering The Latest Research, News, and Information

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Foundation News

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Chiari & Syringomyelia News is published bi-monthly. The next issue will be available November 30th, 2007.

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Conservative Surgery Shows Promise For Adults

The tissue of the brain and spinal cord are covered by three layers, or membranes, which collectively are referred to as the meninges. Working from the brain out, the innermost layer, which is full of blood vessels, is known as pia mater and the middle layer above that is known as the arachnoid.

The sub-arachnoid space (SAS) is the space between the pia mater and the arachnoid through which spinal fluid flows, bathing and cushioning the brain and spine. Finally, above the arachnoid, the dura is the thicker, outermost layer of the meninges.

For years, the dura and the arachnoid have been at the center of a debate in the Chiari surgical community. As this newsletter has documented before, there is a growing trend among pediatric neurosurgeons to preserve either the arachnoid, the dura, or both during Chiari surgery.

These surgeons focus the decompression procedure on removing bone to relieve the pressure and create more space. Some only remove bone and leave the dura completely intact, while others may score the dura or peel it back but leave the underlying arachnoid intact, preserving the CSF space. The advantages of such procedures are that by not penetrating the sub-arachnoid space, the risk of surgical complications are drastically reduced. In addition, the surgery takes less time and patients usually end up going home sooner.

The drawback of not opening the dura is the risk that the decompression will be insufficient to alleviate symptoms or reduce the size of a syrinx. For example, some people have obstructions in the SAS which need to be removed and some surgeons favor reducing or removing the cerebellar tonsils themselves to create enough space.

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Posterior Fossa Volume & Symptoms

Chiari is traditionally defined as the cerebellar tonsils herniating at least 3mm-5mm out of the skull into the spinal area. Although this definition has been used for a long, long time, research in the last 10 years or so has consistently shown that the amount of herniation is not a good indicator of symptom severity or clinical outcome.

In other words, there are many

people with small herniations who have severe symptoms and there are probably even more people with large herniations (greater than 5mm) who have no symptoms. The bad news is that many physicians still rely on the old definition of Chiari, which means that time and again patients with small herniations are told their symptoms are not related to Chiari. *See Page 4*

From The Editor: Setbacks

For those who live with Chiari on a daily basis, setbacks are a way of life. In the nine years since decompression surgery, I have lost count of the number of times I have felt like I was making great strides, only to slip back into the land of pain and inability to do things.

At this point my biggest problem is that the muscles in my neck, upper back and shoulders go into spasm very easily. While this may not sound like much, it can actually be very limiting at times. Besides the near constant pain on the right side of my neck, it can interfere with many daily activities because sitting in most chairs becomes unbearable, as does standing for more than a minute or two. Despite this, I have made great strides over the years through hard work and discipline. In general, today I am able to do things that I never thought I would be able to do again.

In particular, this past summer was going really, really well. I was doing a lot of swimming (which I find helps a lot) and my general activity level was very high. I had started playing guitar again after a 20 year break and quite to my surprise, not only did I retain some skill, but I was able to play without much pain. By the end of the summer I was playing for an hour at a time, something I never thought possible. The peak came on my 40th birthday when I climbed a 50 foot rock wall. Not only did I make it to the top, but I did it easily and with no pain whatsoever.

Then, as has happened so many times before, came the setback. Usually when I get into trouble, and by trouble I mean the muscles in my neck get so still and painful that I can't seem to do anything and nothing helps, it's from a culmination of things as opposed to just one thing. This time it could have been from throwing too many pop-ups to my kids or trying to do one too many push-ups. I've learned not to obsess over what caused it; it almost doesn't matter. The pain and frustration were back and things that I was doing easily just days before were now impossible. I couldn't even hold the guitar without excruciating pain. I was in a place I was unfortunately very familiar with.

In fact, I've been down this road so many times, there are a few things I've learned:

- First and foremost, it's important not to panic. If you're experiencing a setback you have to have faith that things will improve again. If you've already come a long way back, think about how far you've come and how much better your life is than it used to be. I honestly think this is the most important item, everything else is secondary. You have to believe things will get better and over time you will learn not to let

the inevitable setbacks get you down.

- In my experience (and everyone is different), it took a long time for me to learn that the best thing to do was to stop almost everything that might aggravate my neck and take a very conservative approach. As someone who likes to be active, this is very difficult, but once my neck muscles are in spasm they seem to get sparked very easily.
- Fight off the depression any way you can. It's so easy to slip into a funk when symptoms rear their ugly head, but this will only prolong the recovery. Watch a funny movie, indulge in some food, do whatever it takes.
- Once things start to settle down, be disciplined about increasing your activities again. Set a plan and stick with it. If you're like me, then being too aggressive can lead to a downward spiral which you don't want to be in.
- Remember that in order to have a setback, you had to have been making progress. In other words stay positive and stay focused on getting stronger even when you're feeling weak.

In the end, it doesn't matter if your residual problems are headaches, muscle pains, fatigue, or something else. chances are they come and go and that sometimes you feel good and other times you feel bad. So if you're experiencing a setback, keep in mind that the one good thing about setbacks is that, hopefully, they are temporary.

As I work on this issue, it's like there's a knife in the side of my neck. It's almost unbearable, but it's already not as bad as it was a couple of weeks ago. Although the pain is still there, I'm slowly starting to do things again and am heading in the right direction.

--Rick Labuda

Conservative Surgery Shows Promise For Adults

Based upon the UIC/Conquer Chiari Research Symposium 2006, it appears that, at least for children, the concept of preserving the dura/arachnoid is starting to win the debate as more and more pediatric neurosurgeons employ a minimal approach to Chiari surgery. However, to date, even the pediatric proponents of not opening the dura have been hesitant to suggest a similar approach for adults.

A recent study from Italy, however, shows that leaving the arachnoid intact may in fact be a viable surgical alternative for adults as well. The study, Perrini et al., involved 24 adult Chiari patients and was recently published on the Acta Neurochirurgica website.

The Chiari group was comprised of 15 men and 9 women who ranged in age from 25-67 years. They had been suffering for an average of 2.5 years from headaches, sensory disturbances, muscle weakness and other symptoms.

The surgeons employed what they call a conservative approach to surgery. Basically, a minimum amount of bone was removed and while the dura was peeled back and sewn open, every attempt was made to keep the underlying arachnoid intact. It should be noted, however, that in three cases, small pinholes in the arachnoid were made accidentally, and in one case, there was a large tear.

When the patients were evaluated post-surgically, 20 out of 24 (87.5%) showed significant improvement with at least one symptom (see Table 1). Symptom by symptom, the researchers found that all headaches resolved very quickly and the majority of patients had improvements in strength as well. When syrinxes were evaluated on follow-up MRIs, they found that 83% had either collapsed or shrunk and that the rest had stabilized.

Highlighting the increased risks of opening the dura/arachnoid, none of the 20 patients whose arachnoid was intact experienced any complications, but the four whose arachnoids were accidentally penetrated did. Three of the cases were mild, but one person developed hydrocephalus post-surgically and

required a shunt.

Given that past studies have found that not opening the dura/arachnoid in adults results in poorer outcomes, it will take many more patients to determine if this is a valid approach. However, it would be good for the Chiari community to follow the lead of the pediatric surgeons and begin to debate the issue. -- *Rick Labuda*

Source: Perrini P, Benedetto N, Tenenbaum R, Di Lorenzo N. Extra-arachnoidal cranio-cervical decompression for syringomyelia associated with Chiari I malformation in adults: technique assessment. Acta Neurochir (Wien). 2007 Aug 23; [Epub ahead of print]

Table 1

Conservative Surgery Outcomes (24 Adults)

arachnoid - thin membrane covering the

Symptom	% Improved
Headache	100%
Sensory Disturbance	52%
Weakness	83%
Syrinx	83%

brain/spine; lies underneath the dura and above the sub-arachnoid space

cranial nerve - one of a set of nerve pairs which originate in the brain rather than the spine

dura - thicker, outer covering of the brain

sub-arachnoid space (SAS) - space underneath the arachnoid layer through which CSF flows

Key Points

1. Among pediatric neurosurgeons, there is a trend towards leaving the dura, or at least the arachnoid, intact during surgery
2. This lowers the complication rate and speeds recovery
3. Research on such conservative procedures used for adults has been mixed
4. Study looked at outcomes for 24 adult Chiari patients whose surgery left the arachnoid intact
5. Headaches, weakness and syrinx improved in majority of patients
6. Complications occurred only in patients whose arachnoid was accidentally opened during the procedure
7. Provides some evidence that adults may benefit from a limited surgical procedure

Posterior Fossa Volume & Symptoms

The good news is that most, if not all, Chiari experts are aware of the problem and are looking for new ways to define Chiari, objectively measure severity, predict who will develop symptoms and identify good candidates for surgery.

There are several active areas of research in this regard, one of which is morphometric skull analysis. Morphometric is a fancy word which simply refers to the physical size and shape of something, in this case the human skull. Chiari is thought to be a result of the underdevelopment of certain parts of the skull (which are then too small for the brain) and researchers have begun to use MRIs to quantitatively measure skull dimensions.

Studies using this technique have shown that on average Chiari patients have small posterior fossas as compared to healthy people. Research has also shown that skull size may be linked to the development of a syrinx and that people with very small herniations, but Chiari-like symptoms, tend to have abnormal skulls as well.

Now, in a study posted in July on the Child's Nervous System website, a group from Canada (Trigylidas et al.) revealed that even some asymptomatic people with herniations have abnormally small posterior fossas. The Canadian team looked at the skull shapes of 61 pediatric Chiari cases from the Eastern Ontario Children's Hospital and compared them to 20 controls who had no history of intracranial problems.

In the Chiari group, 34 children were considered symptomatic with headaches, scoliosis, balance problems, and muscle and sensory problems being the most common. To assess the size of the posterior fossa, the researchers used MRIs to measure the total intracranial volume (ICV) and the volume of the posterior fossa (PFV). They then calculated the ration of PFV to ICV as a measure of how large the posterior fossa is relative to the total skull. This ratio method is a technique other researchers have used to account for the variability in skull sizes of children of different ages.

Not surprisingly, the data showed that on average the Chiari group (both symptomatic and asymptomatic) had a smaller PFV/ICV ratio than the control group (see Table 1). However, the Canadian researchers went further and analyzed the data by comparing symptomatic to asymptomatic and by age. Since the average age of the Chiari group was 10, the scientists split the data into two group, 0-9 years and 10-18 years.

The results of this analysis were intriguing. In the younger group (0-9), both asymptomatic and

symptomatic Chiari patients had smaller PFV/ICV ratios. However, in the older group (10-18) while the symptomatic children again had small ratios, the asymptomatic children had normal PFV/ICV ratios. In other words among the asymptomatic children, the younger ones had abnormal skull shapes, while the older ones had normal skull shapes.

Based on this, one has to wonder if some young children with small posterior fossas become symptomatic in time, while in others the skull growth catches up and they remain symptom free. What would be really useful is a longitudinal study which identified young children with asymptomatic herniations and small posterior fossas and followed them over time as they grew up.

As an interesting side note, the researchers also looked at CSF flow data for a subset of the children. They found that both symptomatic and asymptomatic children demonstrated abnormal flow characteristics. While some surgeons have begun to rely on flow studies, others have questioned their utility and this result explains why.

It appears that what makes people with herniations (even small ones) symptomatic is either very complicated or due to something that for the time being remains unknown. What is clear is that identifying that unknown would not only provide a way to objectively measure Chiari, but may lead to a redefinition of the entire problem.

-- Rick Labuda

Source: Trigylidas T, Baronia B, Vassilyadi M, Ventureyra EC. Posterior fossa dimension and volume estimates in pediatric patients with Chiari I malformations. *Childs Nerv Syst.* 2007 Jul 27; [Epub ahead of print]

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Posterior Fossa Volume & Symptoms

Table 1
PFV/ICV Ratio By Age and Symptomology

Age	Asymptom	Symptom	Control
0-9	.105	.103	.127
10-18	.118	.106	.127
Total	.115	.103	.127

Note: PFV = posterior fossa volume; ICV = intracranial volume; asymptom = no symptoms; symptom = symptoms attributable to Chiari

Related C&S News Articles:

Small Posterior Fossa Linked To Chiari Related Syringomyelia

Is Chiari 0 For Real? Research Shows Small Posterior Fossa In Chiari 0 Patients

Chiari Link To Small Posterior Fossa Confirmed In Adults

CSF Flow In Children Before & After Surgery

asymptomatic - not having any symptoms

herniation - the protrusion of a body part into an area where it isn't normally located

intracranial volume (ICV) - a measure of the total amount of space inside the skull

morphometric - referring to the physical structure and size of something, in this case the skull

posterior fossa - region in the lower back part of the skull where the cerebellum is situated

posterior fossa volume (PFV) - measure of the total amount of space occupied by the posterior fossa region

PFV/ICV ratio - measure of how much of the entire skull the posterior fossa takes up; used as a way to eliminate the variability of skull sizes in children of different ages

tonsillar herniation - refers to the cerebellar tonsils extending below, and out of, the skull into the spinal area

Key Points

1. Research has shown that the classic definition of Chiari, based on length of herniation, is not very useful
2. Based on the theory that Chiari results from a small posterior fossa, one area of interest is measuring posterior fossa volume and skull dimensions
3. Previous research has found that Chiari patients tend to have smaller posterior fossa regions than normal
4. This study looked at 61 pediatric Chiari patients as compared to normal controls
5. Found that overall Chiari group had smaller posterior fossa as measured by the PFV/ICV ratio
6. Also found that for children age 0-9, both symptomatic and asymptomatic subjects had small posterior fossas
7. However asymptomatic older children had skulls similar to the controls
8. Long-term studies would be required to see if skull size/shape is useful for predicting who will develop symptoms

Complex Chiari Cases Have Poorer Outcomes

Complex Chiari cases, defined as the existence of additional abnormalities such as fused vertebra or basilar invagination, have poorer surgical outcomes than simple Chiari cases. That was one of the findings from a study published recently in the journal, *Pediatric Neurosurgery*.

The study, from a group of doctors in China (Guo et al.), reported on the surgical outcomes of 128 Chiari cases treated over several years at Zhengzhou University. At this point there have been several surgical outcome reports involving a large number of patients, which has pretty much established that about 80% of patients experience at least some type of relief as a result of surgery. However, the Chinese team went a step further and separated their patients into what they called simple and complex cases.

Their study involved 128 consecutive patients, with MRI confirmed Chiari of at least 5mm, who underwent decompression surgery. Most of the patients were adults (112) and the majority had Chiari I (115). Surprisingly, 95% of the large group also had at least one syrinx. As to be expected, there were a variety of symptoms, with motor weakness and temperature and touch sensory disturbances being the most common (see Table 1).

The surgery was tailored to each patient's individual needs, but generally involved a craniectomy, laminectomy and duraplasty, with removal of the cerebellar tonsils as needed. In addition, 28 patients required fusion of the vertebra with an autogenous bone graft and 5 required stabilization with plates and screws.

For the purposes of analysis, outcomes were categorized as resolved, improved, unchanged and worsened, with resolved or improved considered a good outcome and unchanged or worsened a poor outcome. Patients were evaluated immediately after surgery and for an average of three years post-operatively, although 24 patients did not return for long term follow-up.

Overall, outcomes were good with 81% of the pediatric patients and 90% of the adults showing improvement in symptoms immediately after surgery. In addition, post-surgical MRIs showed that syrinxes collapsed or shrank in 79% of the patients with SM.

To look deeper into the data, the researchers next categorized all the patients as having either a simple case of Chiari or a complex one. Complex cases were defined as having one or more additional abnormalities, such as congenital fu-

sion of the top two vertebrae or basilar invagination. Using this definition, there were 90 simple cases (70%) and 38 complex ones (30%).

When they broke the data up this way, they found a significant difference between the outcomes for the simple and complex cases. Specifically, while an impressive 95% of the simple cases had good outcomes, only 74% of the complex cases did (see Table 2). As an extension of this, the surgeons also found that patients who underwent fusion as part of their surgery were significantly more likely to have a poor outcome. This makes sense, seeing as how many of the structural problems which define a complex Chiari case may require fusion to provide stability. And while it seems intuitive that complex cases may not fare as well, it is very useful to see the data broken out this way to quantify it.

Long term follow-up averaged 3 years and showed that overall there was complete resolution of symptoms for 28% of the patients, while an additional 59% experienced significant improvement. Many studies would combine these two categories as a measure of success, which would produce a pretty impressive 87% success rate for the surgery.

While the overall success rate was high, there was also an unusually high rate of complications. There were no mortalities associated with the surgery, but 28% of the patients did experience complications such as fever, CSF leak and instability.

There are now enough surgical outcome reports such as this one to say that decompression surgery improves symptoms for a majority of patients. However, since this type of report is one of the most common in the Chiari literature, it would be useful if surgeons began to dig deeper into how they classify patients and outcomes. This study represented a good step in that direction by looking at simple versus complex cases and breaking outcomes into four distinct categories.

- Rick Labuda

Source: Guo F, Wang M, Long J, Wang H, Sun H, Yang B, Song L. Surgical management of Chiari malformation: analysis of 128 cases. *Pediatr Neurosurg.* 2007;43(5):375-81.

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Complex Chiari Cases Have Poorer Outcomes

Table 1
Common Symptoms (128 Chiari Cases)

Symptom	% With
Motor Weakness	60
Loss of Temperature Sensing	59
Loss of Touch Sense	58
Headache	35
Neck Pain	30
Limited Neck Motion	46

Table 2
Surgical Outcomes, Simple vs Complex Chiari Cases

	Good	Poor
Simple (90)	95%	5%
Complex (38)	74%	26%

Note: Complex refers to Chiari in combination with other abnormalities, such as congenital fusion or basilar invagination; Good = resolved or improved; Poor = unchanged or worsened

Table 3
Long-Term Surgical Outcomes (104 Cases)

	Pediatric (15)	Adult (89)
Resolved	4	25
Improved	8	53
Unchanged	2	6
Worsened	1	5

arachnoid - thin, middle layer of the coverings of the brain and spinal cord

autogenous - taken from the patient's own body

central canal - tube like center of the spinal cord; usually collapses as people age

Chiari II - more serious form of Chiari; associated with spina bifida

craniectomy - surgical technique where a piece of the cranium, or skull, is removed

dura - thick, outer covering of the brain/spinal cord

duraplasty - surgical technique where the dura is expanded with a patch or graft

laminectomy - surgical technique where part of one or more bony vertebra are removed

mortality - death rate

obex - opening at the top of the central canal of the spinal cord

vertebra - one of the bony segments of the spine

Key Points

1. There have been several surgical reports involving large numbers of patients which have shown about an 80% improvement rate after surgery
2. Study from China reported surgical outcomes for 128 patients
3. Overall success rate was good; researchers separated cases into simple and complex
4. Simple cases had good outcomes 95% of the time, while complex ones only had good outcomes 74% of the time
5. Of all the cases that had poor outcomes, patients who required some type of fusion for stability had poorer outcomes

Extensive Laminectomy May Increase Risk Of Spine Problems

Scoliosis, an abnormal curvature of the spine is widely known to be linked with Chiari and syringomyelia, yet the exact nature of the relationship is not clear. Research has shown that CM/SM patient tend to have abnormal curve patterns, even for scoliosis, and that decompression surgery often stops the progression of scoliosis in children. However, attempts to correlate syrinx size and location to the presence and severity of curves have failed. Despite this, it is still believed that syrinxes affect certain nerves in the spinal cord leaving people vulnerable to spinal problems such as scoliosis.

Now, a report from an experienced surgeon, Dr. Ulrich Batzdorf, in the August, 2007 issue of Neurosurgery (Batzdorf et al.) indicates that certain aspects of surgery to treat Chiari and syringomyelia may also contribute to spinal problems. In the study, Batzdorf reviewed 169 syringomyelia cases he had treated operatively over the previous 20 years.

Most of the patients had Chiari related syringomyelia (105) while the rest had primary SM due to trauma, tumors and meningitis. In the CM/SM group sixty-three of the patients were treated with an initial operation, while 42 had already had some type of decompression surgery and Batzdorf was performing a reoperation. In all cases MRIs were reviewed for signs of spinal abnormalities such as scoliosis and kyphosis.

Interestingly, the majority of patients in the first time surgery group had no significant spinal abnormality (58%), while the opposite was true for the reoperation group (see Table 1). Specifically, 57% of the reoperation group demonstrated significant spinal abnormalities. In addition, even when scoliosis was present in the first time group, it was usually mild, whereas the scoliosis found in the reoperation group was mostly classified as severe.

In comparing patients who did and did not have significant spinal abnormalities, the researchers found a major difference involving the amount of bone removed during laminectomies. Recall that a laminectomy is a procedure where part of one or more bony vertebra is removed. Usually a Chiari decompression will involve a laminectomy starting at the top vertebra and going down as far as the cerebellar tonsils descend. In addition, laminectomies are sometimes performed at the level of a syrinx to locally decompress the area or allow for the placement of a shunt to drain the syrinx. In this study the aver-

age amount of bone removal for patients with no spinal deformity averaged 2.6 vertebral segments. However, this jumped to 4.7 segments for the group with spinal deformities.

To assess long term outcomes, the researchers attempted to contact each patient and were able to ascertain the status of 163 out of the original 169. The contact was brief and simply designed to determine each person's status relative to how they were immediately after recuperating from surgery. Not surprisingly, the majority of patients reported they felt about the same, but there was a significant difference between the first time operation group and the reoperation group. Specifically, only 3 out of 62 people in the first time group reported being worse off, whereas 9 of the 41 people in the reoperation group were worse.

After looking at the data further, the researchers identified a subset of six patients whose severe spinal deformities coincided with the time and location of surgical procedures. Based upon all the results, Batzdorf believes that laminectomies beyond the top cervical vertebra (C1) increase the chance of spinal problems like scoliosis or kyphosis developing or becoming severe. Basically, the combination of nerve damage from a syrinx, combined with the structural changes from a laminectomy can weaken things to the point that problems develop.

Because of this, the authors recommend that if bone needs to be removed beyond the top vertebra (C1) that a modified technique be used which can preserve musculature and a key ligament. In comments published in the same journal issue, several surgeons agreed with Batzdorf's approach, although a couple did point out some problems with the design of the study and stressed the need for more research in this area.

- Rick Labuda

Source: Batzdorf U, Khoo LT, McArthur DL. Observations on spine deformity and syringomyelia. Neurosurgery. 2007 Aug;61(2):370-7

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Extensive Laminectomy May Increase Risk Of Spine Problems

Table 1
Percent of Patients With Spine Deformity (105 Total)

	First Chiari Surgery (63)	Chiari Reop. (42)
No Significant Deformity	58%	43%
Significant Deformity	41%	57%

Table 2
Average Laminectomy Size (Vertebral Segments) No Deformity vs Deformity

	Laminectomy Size
No Spinal Deformity	2.6
Significant Spinal Deformity	4.7

Note: Laminectomy size is measured in vertebral segments

Table 3
Long Term Outcome Status, First Surgery vs Reoperation

	Better	Same	Worse
Initial Surgery (62)	18	41	3
Reop. (41)	5	27	9

Note: Status is as compared to discharge after surgery; number of patients in each group is represented

cervical - upper part of the spine, neck area

kyphosis - abnormal forward curvature of the spine which effectively creates a hump

laminectomy - surgical technique where part of one or more bony vertebrae are removed

meningitis - infection or inflammation of the meninges, the layers which cover the brain and spine

scoliosis - abnormal spinal curvature

thoracic - the middle part of the spine, chest area

vertebra - one of the individual bony segments of the spine; referred to by region and number, for example C1 is the first segment in the cervical region

Key Points

1. Scoliosis is linked in an unknown way to CM/SM
2. Research has shown that CM/SM patients have unusual curve patterns even for scoliosis; however the size and location of syrinx is not related to curve severity
3. Batzdorf reviewed 169 SM surgery patients over a 20 year period
4. Found that patients undergoing a second operation were more likely to have a spinal deformity, such as scoliosis, than first time patients
5. Deformities also tended to be more severe among reoperations
6. Also found that the average laminectomy size was larger among the reop group and long term outcomes were not as good
7. Author believes that extensive laminectomies (below C1) combined with a syrinx leave people prone to spinal problems
8. Recommends a modified laminectomy lower on the spine

New Dural Patch Found To Be Safe & Effective

While the debate over whether it is even necessary to open the dura as part of a Chiari decompression rages on, advances in the types of materials that can be used for dural grafts continues. In fact, today's surgeons, and patients, have an array of dural patch options, including:

From the Patient – Many surgeons have begun to use tissue from the patient's own body for the dural graft. The tissue can be taken from a number of different places and has the advantage of eliminating any type of immune response to the graft. Some people also believe that this type of graft reduces the chance of infection. The downside, of course, is that there is another surgical site which needs to heal and may cause additional pain.

Cadaver – Very popular a number of years ago, grafts from cadavers are being used less today because of safety concerns. Cadaver grafts can be made either from dural tissue directly or the pericardium, which is the membrane around the heart.

Cow – Bovine grafts, such as DuraGuard (Synovis), are taken from the animal's pericardium.

Collagen – Several manufacturers have recently come out with a new class of products known as collagen matrices. Collagen is a type of connective tissue which provides structure to body parts and has several advantages as the basis for a graft. Two of these grafts, Durasis (Cook Biotech) and DuraGen (Integra Lifesciences) both utilize animal collagen to form pliable, easy to work with grafts, which are actually absorbed by the body's tissue over a short period of time.

Synthetic – Dural grafts can also be made from completely synthetic materials, such as Gore-Tex.

With so many options, what do surgeons look for when they choose a dural graft? An ideal dural substitute must be both safe and effective, meaning that it should be pliable, easy to suture, able to hold a watertight seal to prevent CSF leaks, integrate quickly into the natural tissue of the body and result in minimal complications. While this sounds like a tall order, a publication in the June, 2007 issue of the *Journal of Neurosurgery* by Dr. Bejjani of the University of Pittsburgh and Dr.

Zabramski of the Barrow Neurological Institute indicates that a newer graft, Durasis is a strong candidate.

Bejjani and Zabramski reported the results of a multi-center trial of the dural substitute in 59 patients treated for a variety of reasons, including Chiari, tumors, and tethered cords (see Table 1). According to Cook Biotech, the manufacturer, Durasis "utilizes Cook's innovative Small Intestinal Submucosa (SIS) technology, a collagen biomatrix that supports regrowth of host tissue. SIS is a natural biomaterial harvested from pig small intestine and developed into strong, sterile, pliable sheets that provide a rich environment for cell attachment and tissue growth. Durasis takes on the cellular characteristics of surrounding dural tissue while it serves as a scaffold on which human tissue grows. As the biomaterial is replaced with cells, the scaffold is no longer needed and it is absorbed by the body. The result is a tissue that looks and functions very much like the tissue it replaced."

To evaluate the safety and efficacy of Durasis, surgeons at several centers carefully tracked its use and rated its effectiveness on a 5 point scale, ease of use on a 4 point scale, and looked for complications due to the graft at several points in time after surgery.

For the 59 patients in the study, the overall complication rate was fairly low with the only problems being 1 CSF leaks (1.7%) and 2 infections (3.4%). To compare this with other materials, the authors selected 10 published studies which reported complications from a variety of dural graft types and comprised over 1800 patients. The combined complication rates from the 10 studies was 5.2% for CSF leaks and 5.0% for infections. It should be noted that there no CSF leaks for the 32 Chiari surgeries using Durasis.

In terms of efficacy, Durasis scored an average of 4.8 on a 5 point scale and the graft was considered successful in all 59 cases. In addition, the surgeons rated the material as excellent in handling issues, such as strength and ease of suturing.

While these results indicate that Durasis appears to be a good dural patch, it is important to keep in mind that there are other good options as well and not every surgeon is going to switch to using Durasis. It is also important to keep in mind that this study was funded by Cook Biotech as part of the FDA approval process.

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New Dural Patch Found To Be Safe & Effective

In fact, research presented at the 2007 UIC/Conquer Chiari Research Symposium by Dr. Konstantin Slavin (UIC) indicated that it may not matter what material is used. Dr. Slavin presented the results of a randomized prospective study of 25 patients which compared two well-known graft products. One of the grafts required sutures to be put into place, while the other one did not.

The grafts were evaluated on clinical outcome, complication rates, length of hospital stay and time in surgery. Interestingly, there was no difference between the two grafts for outcome, complication rates, or length of hospital stay. The good news is that today's Chiari surgeons and patients have more choices for dural grafts than in the past and that their safety and effectiveness is improving.

- Rick Labuda

Source: Bejjani GK, Zabramski J; Durasis Study Group. Safety and efficacy of the porcine small intestinal submucosa dural substitute: results of a prospective multicenter study and literature review. J Neurosurg. 2007 Jun;106(6):1028-33.

Table 1
Surgical Procedures Used In Study (59 Total Patients)

Procedure	# of Patients
Chiari I	32
Tumor	18
Tethered Cord	3
Aneurysm	3
Other	3

Table 2

Complication Rates, Durasis vs Medical Literature Reports

	Durasis	Literature
CSF Leak	1.7%	5.2%
Infection	3.4%	5.0%

Note: The Literature rates comprise the combined complication rates of 10 published studies of various graft materials (selected by the authors) involving over 1800 patients

aneurysm - a bulge in the wall of an artery due to an inherent weakness

collagen - found in connective tissue and acts as an important support structure for cells

CSF leak - complication where CSF is not totally enclosed beneath the dura and leaks out

dura - thicker, outer covering of the brain/spinal cord

dural patch - material, such as from a cow, collagen, or the patient which is sewn into an opening in the dura

duraplasty - surgical technique where a patch is sewn into the dura

Key Points

1. Several types of dural graft materials are available today, including grafts taken from the patient, cadavers, cows, collagen, and synthetic
2. Multi-center study looked at the safety and efficacy of 59 patients using Durasis graft
3. Found very favorable complication rates, including no CSF leaks in 32 Chiari surgeries
4. Efficacy of graft averaged a score of 4.8 out of 5 and was considered successful in every patient
5. Surgeons rated the handling of the material as excellent
6. Research presented at CC Symposium showed that graft choice may be a matter of surgeon preference

Ray's Corner: Everyone Needs A Hug

[Ed. Note: The opinions expressed below are solely those of the author. They do not represent the opinions of the editor, publisher, or this publication. The author is not a medical doctor and does not give medical advice. Anyone with a medical problem should seek professional medical advice]

Everyone needs a hug.

I usually write about technical or semi-technical topics. I thought I would do something different this time and write about the human side of Chiari. Let my female side show a little, just a little. I want to talk about what it is like to have Chiari at the emotional level and why everyone with Chiari needs support and genuine understanding especially from those closest to them.

Chiari was particularly aggressive with me. It more or less started with pain upon swallowing probably due to compression of the lower cranial nerves and 18 months later reduced me to using a wheel chair. I had strength in my legs but when I attempted to use it, it made me sick. It was also true of my arms but even worse so I couldn't even push my own wheel chair. I had many other symptoms as well; too long a list to reconstitute here.

I was on disability. I pretty much came to the conclusion that I would never feel well again. I was depressed and sometimes thought about suicide. The doctors were convinced that clinical depression was my problem and the cause of my physical symptoms. They had pretty much convinced my wife which really drove me into loneliness. When your spouse is forced into a state of confusion, you are truly alone.

I was the only one supporting my family. My wife did not work. I worried about my family on many levels. How would I support them? What if I became a burden to them? How was I going to raise my son and be there for him? I worried about gradually declining which would lead to signifi-

cantly shortening my life span.

The worry was absolutely overwhelming. Doctors could not tell me what was wrong and didn't particularly care to find out. They just wanted me to continue talking antidepressants and seeing the psychiatrist. When you know clinical depression is not part of your normal make up and no one believes you, you feel completely helpless. Not getting a diagnosis drains you of hope and living in a situation where no one is the least bit curious about finding a diagnosis is a nightmare.

Decompression is fairly painful particularly the first couple of weeks after surgery. Your head feels like it will explode with the slightest movement. You have to stay in bed most of the time the first couple of weeks after coming home. You lay in bed in such a great deal of pain that you wonder if you made the wrong decision in having surgery. Most surgeons will only promise that the surgery will halt the progression of the syndrome but will not promise improvement. You lay there and have serious doubts that you will experience improvement.

My parents drove 600 miles to be there for me during and after my surgery. They were old and had their own problems but there is no way to stop parents when their child is in need.

The first day I was home after the operation, my mother was at my side in my bedroom. She looked at me with the back of my head all bandaged up and could feel my pain. Tears came to her eyes and she began to weep. She bent down and hugged me and told me that she wished we could trade places. Even though I was 48 at the time, I needed to hear that and when I did, I too cried because I realized just how deep her love was for me. I assured her that I would fight back. Her display of love at that very moment was an important turning point for me. Even though I was the one suffering, I could not bear to see her sadness. I wanted her to be happy and I resolved to do my best to recover.

My mother passed away a couple of years later

Ray D'Alonzo, Ph.D., is Manager of Doctoral Recruiting & University Relations and a former Associate Director of Research and Development at Procter & Gamble Pharmaceuticals where he has worked for 30 years. He has led research programs in bone metabolism, infectious disease, respiratory disease, arthritis, and nutrition and has published scientific papers on a wide variety of topics from the chemical composition of fats and oils to the pharmacoeconomics of osteoporosis. Dr. D'Alonzo is the recipient of the Chancellor's Medal from the University of Massachusetts, Amherst, in part, for his contributions to the development of new pharmaceutical agents. As both a patient and scientist, he has made a personal effort to increase the awareness of Chiari in the health care sector and to assist others afflicted with the syndrome. He has published the story of his personal struggle with Chiari in a book, [Contents Under Pressure](#), with 100% of royalties going towards Chiari education, awareness, and research programs.

Ray's Corner: Everyone Needs A Hug

from idiopathic pulmonary fibrosis. She suffered far greater than I during the last two years of her life. At one point she developed pneumonia and was hospitalized. She had glaucoma and while in the hospital she contracted an eye infection from a nurse or aid who administered her eye drops. The infection could not be controlled and she had to have her eye removed a couple of months later. The eye she lost was her good eye and she was left blind for the last year of her life while she was anchored to an oxygen tank at all times.

I was at her side in the hospice for her last two days. I watched her struggle to take every breath. I kept her lips moist and put ice chips in her mouth. I, my father and brother stayed with her to the end. Our hearts were ripped out at the end but we all felt relieved that she was out of her misery. She remained true right up to the moment that she went unconscious. She never despaired and her mind was clear. She was brave and good and full of love.

Whenever I think about her, I remember her crying over my bedside and pleading to trade places with me. All the times she disciplined me or nagged me, I really recall not. I remember her love most of all, the love that gave me resolve to fight back.

Today, almost 8 years later, I am better than ever. In many ways, I have never felt so good. Yes, it was the surgeon's knife that played a key role in giving my life back but without my mother's loving hug, I wonder if the ultimate outcome would have been as effective.

Postscript - I recently learned of the death of a great woman, Dr. Marcy Speer. I never had the privilege of meeting her but I am familiar with her work in the genetics of Chiari. Her paper in the Journal of Genetic Counseling, volume 12, No. 4, August 2003, is a paper I have referenced dozens of times as the best review article on the prevalence of Chiari. Marcy lost her battle against breast cancer on August 4th but her contributions to Chiari research constitute a significant victorious battle in the war against Chiari. -- Ray D'Alonzo

**** If you would like to share your comments, thoughts, or ideas with Ray, please send them to dalonzo.rp@fuse.net. Due to the volume and nature of email received, individual responses are not possible. ****

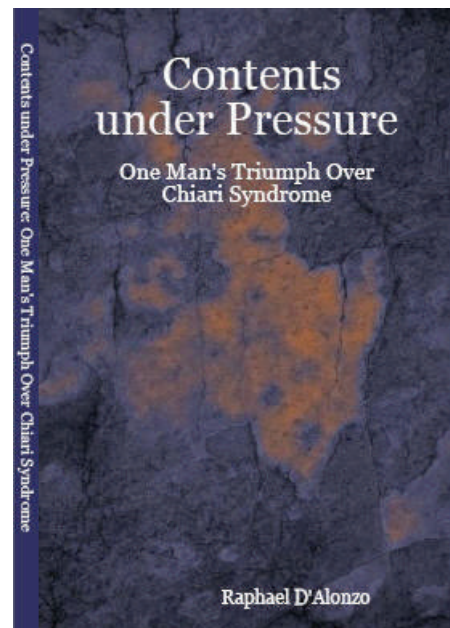
Contents Under Pressure, By Ray D'Alonzo

You may have cheered for him when he ran the Air Force Marathon to raise money for Conquer Chiari, now you can read his whole, amazing story. From wheelchair to marathon, Ray's story will inform, inspire, and raise money to fight Chiari.

Learn More Or Purchase This Book:

[Contents Under Pressure](#)

(Note, this book is offered through www.lulu.com. Clicking on this link will take you off the Conquer Chiari website. Conquer Chiari is not responsible for the content of the site linked above.)



"In publishing this book, I am trying first and foremost to increase the general awareness of Chiari. All too often, Chiari is unrecognized, misdiagnosed or dismissed. Second, Chiari patients who read this book will undoubtedly validate many of their own symptoms and come to realize that they are not alone nor crazy. Third, patients must understand that recovery from decompression surgery and the syndrome itself is often very protracted but with time, the body's remarkable ability to adapt and repair itself, and a little self perseverance, hope exists for many (unfortunately, not all) who continue to suffer. Finally, 100% of all the royalties from this book are paid directly to the Conquer Chiari campaign to support research and patient/health care provider education programs."

In The Spotlight: Dr. Raymond Sekula, Neurosurgeon

Dr. Raymond Sekula is a neurosurgeon at Allegheny General Hospital in Pittsburgh. Dr. Sekula has treated many Chiari patients and has published on the subject. When he approached Conquer Chiari saying he wanted to help our cause, we of course took him up on his offer.

As a first step, we put Dr. Sekula In The Spotlight...

About how many Chiari patients do you see each year?

I see more than 150 patients referred with "Chiari malformation" each year.

Surveys have shown there is a lack of agreement in the surgical community on when surgery is required, what is your criteria for recommending surgery?

There is a lack of agreement in the neurosurgical community because many surgeons are uncomfortable with Chiari malformation. Ignorance in this area persists. In time, surgeons will acknowledge the heterogeneity of complaints referable to Chiari malformation and begin to treat patients rather than imaging studies.

Approximately what percent of patients that you evaluate end up having surgery?

Approximately 50%.

There is also a great deal of variation among surgeons regarding the specifics of the Chiari surgery, how would you describe your standard surgery?

I believe that there are multiple procedures appropriate for patients with Chiari I malformation. Some may do well with bony reduction only while others may do well with tonsillar reduction alone. My standard surgery involves a one inch incision -within the hairline - with a small amount of bone removal (preventing later "cerebellar sag"), bilateral tonsillar reduction with or without a duraplasty.

What are the advantages of performing the surgery in this way?

Ease of postoperative recovery, better cosmetics, and I believe improved long-term outcomes.

How do you define a successful surgery, and what percent of patients with Chiari only (no syrinx) have a favorable outcome?

A successful surgery is one in which a patient obtains marked improvement in symptoms if symptomatic. If a patient is asymptomatic, radiographic improvement (ie. resolution of syrinx) is critical. More than 90% of patients without a syrinx will have improvement in their symptoms.

Can patients with a syrinx ever expect to be symptom free?

For patients with syringomyelia, resolution of symptoms can be expected if surgery is performed early in the course of the disease, for example a patient with a few years of symptoms.

What advice do you give patients on recovering from surgery, returning to work, etc.? Do you recommend physical therapy after decompression surgery?

My patients spend the night after surgery on the regular nursing floor, and I discharge them on the first or second morning following surgery. They can wash the hair and incision on the third postoperative morning. They may return to work in three weeks. I do not recommend physical therapy for fit individuals.

Many people are now describing Chiari as a problem with the size of the posterior fossa, do you agree with this? Do you think this applies to most/all Chiari patients or only a subset of cases?

Marin-Padilla published an influential report in 1981 entitled "Morphogenesis of the experimentally induced Arnold-Chiari malformation." In that report, they induced a small posterior fossa with vitamin A and caused a Chiari malformation.

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In The Spotlight: Dr. Raymond Sekula, Neurosurgeon

You published a paper which showed that patients with minimal herniation, but with Chiari-like symptoms, had similar skull dimensions to Chiari patients with larger tonsillar herniations. Do you think this validates the concept of Chiari 0?

Well, maybe. Jerry Oakes published an interesting paper entitled "Analysis of the posterior fossa in children with the Chiari 0 malformation" which indicated these patients have a smaller posterior fossa. In addition to only evaluating six patients, there were other shortcomings to the report, as there were with our morphometric paper.

Research has shown that the traditional definition of Chiari is not really that good, do you think Chiari should be redefined? If so, how?

Absolutely. Tonsillar descent is only one indicator of a posterior fossa problem. This will be resolved in the next decade and the key is morphometric analysis of the posterior fossa.

Why do you think some people develop syrinxes and others don't?

I cannot answer this.

The Piston Theory states that the movement of the cerebellar tonsils, as driven by the cardiac cycle, creates a pressure wave of CSF which forces the fluid into the spinal tissue to form a syrinx. Do you agree with this theory?

I like this theory, but it is a theory - this is one reason I feel it is necessary to perform a tonsillar reduction rather than a large duraplasty. I prefer to treat directly rather than indirectly.

Are you working on any research at this time that you can discuss?

Yes, I am working on posterior fossa morphometrics and outcome analysis.

How do you see Chiari surgery evolving over the next 5-10 years and do you think there will ever be a non-surgical alternative for treatment?

Although I am a surgeon, I would very much like to see a non-surgical alternative for Chiari I malformation. If Marin-Padillas' mesodermal underdevelopment theory is validated in the future, perhaps we can make a difference at the developmental stage during pregnancy as folic acid has for neural tube defects.

Selected Publication:

Sekula RF, Jannetta PJ, Casey KF, Marchan EM, Sekula LK, McCrady CS.
Dimensions of the posterior fossa in patients symptomatic for Chiari I malformation but without cerebellar tonsillar descent. *Cerebrospinal Fluid Research* 2005, 2:11 (18 Dec 2005)

Good News Story: College Student, Competitive Swimmer, Chiari Conqueror

Kelly deMarrais is a student at Virginia Tech, a competitive swimmer and a great Chiari story...

How would you describe yourself before you became affected by Chiari? In other words, were you physically active, etc.

Before I became affected by Chiari, I was very active with sports. I was playing softball, basketball, field hockey, and swimming. I loved being on a team and competing.

When did you first notice symptoms and what were they?

I first noticed symptoms after I came home from a 7th grade school dance towards the end of the school year. I had to lay down because my head was hurting so badly, but I thought it was just from dancing. The head aches/head pains that I was getting only occurred once a week in the beginning. But as time passed and I was heading into 8th grade, I soon realized that these pains were abnormal and weren't caused by doing any certain activity. As more time passed, I got more symptoms, before I knew it, my hands were going numb for no reason, my feet and fingers felt like pins and needles, and shooting head pains occurred almost every minute.

Were you diagnosed quickly, or did it take awhile?

I was diagnosed rather quickly. I went to my doctor at the beginning of 8th grade and told him that I was having head pains for no reason and he scheduled me for an MRI. I found out right away that I had Chiari malformation.

Was the decision to have surgery easy for you, or were you unsure about it?

The decision to have surgery was probably one of the hardest decisions my family and I had to make. I was not sure how much the surgery would work, if it would even help, or how it would affect my swimming career and my life. It took me over a year and a half to finally decide to get the surgery. I wanted to wait till I had a break with swimming and wouldn't have to worry about missing any school work.

What did your doctor tell you about whether you'd be able to return to competitive sports?

The doctor told me that if all went well that I would be able to return to swimming in no time. He said that swimming would actually help in my recovery as well, because it would help me build my neck muscles and rotation back up.

What was your recovery from surgery like?

Recovery from surgery took a little longer than I expected. I had never had any kind of surgery before, so I didn't have much of an idea what it would be like. It took 2 full months for all my symptoms and everything to go away. I had to do a lot of therapy to work on building up strength to be able to move my head, and rotate my head in both directions. But, once all my symptoms fully went away, I felt better than ever. It was weird not getting head pains, or having my hand and feet have pins and needles, I had always thought that that would have to be something that I would just have to live with for the rest of my life.

Can you talk a little bit about your swimming?

I have been swimming competitively since the age of 6 and have been swimming all year round (have a total of basically 1 month off a year) since the age of 8. I qualified for YMCA Nationals at the age of 12 and attended that meet until I was a senior in High School. The summer before going to college, I was a YMCA National Champion in the 200 Long Course Meter Backstroke. I swam for Somerset Valley YMCA in NJ and currently swim at Virginia Tech. I am the current record holder at Virginia Tech in the 200 yard backstroke.

Did you always feel like you'd be able to go back to it?

I was quite hesitant as to whether or not I'd be able to get back into swimming, especially at the level that I had been competing at prior to surgery. I was not able to dive into the water until 3 months after surgery. But I achieved all best times in most of my best events after only 4 months after having surgery, that was something that I never expected. I had hoped that I would go within a few seconds of my best times, I never imagined dropping time.

How do you feel now, are you 100%, and how is the swimming going?

I definitely feel 100% now, I cannot tell you how much surgery has truly helped me with not only swimming and athletics, but being able to enjoy life as well. My doctor truly changed my life for the better. I now am able to live pain free. Swimming is going well, I am currently trying to make an Olympic Trial cut in the 200 Long Course Meter Backstroke, I am only 8 tenths off the cut.

What would you say to other Chiari people facing the prospect of surgery?

I think surgery is definitely worth getting. It changed my life in so many ways.

Fundraising: Mini Golf Tournament Great Way To Raise Money

Ed. Note: *Because of the importance of raising money for research, the next few Newsletter issues will contain an article with tips on how to organize a specific type of fundraiser. If you have ideas for this column, or would like to write one, please contact director@conquerchiari.org.*

Many people would like to organize a fundraiser but worry about whether they have the time and energy to actually do it. One idea for a fundraiser that can be small and easy, or as large and involved as you want it to be, is a Miniature Golf Tournament. Unlike a large charity golf outing at a country club, which can take a lot of time and effort to organize, a Miniature Golf Fundraiser is more manageable, but can still be a very worthwhile event.

Note, some of the tips below are taken from an article by Kimberly Reynolds:

Tips For Organizing A Miniature Golf Event

- Just like with any event, start planning early. The earlier you start, the easier it will be to get organized and the more time you will have to line up sponsors, publicity, etc.
- Use the Conquer Chiari Meeting Place to find other Chiari people in your area that might be interested in helping. Fundraisers which bring together different networks of people are usually more successful.
- Pick a venue and a date. Call local mini golf establishments and ask how the fee would work (flat rate for a certain amount of time or per player). Also, establish what the rules would be for hanging signs (from sponsors), having food there, etc. Take into account whether the course has a room where an after event could be held or is near a park where there could be a cookout. In picking the date, consider what the weather is likely to be.
- Create a flyer for the event. The flyer should give the specifics of the event, some general info on Chiari, what the money that is raised will be used for (Chiari research through Conquer Chiari, for example), how to register, and who to contact for more information. If you're selling tickets to the event (likely the main way you're raising money), it is important to print directly on the ticket how much of the ticket price is considered a donation. For example, if the facility is charging \$5 per person to play and you decide the ticket price should be \$10 per person, then \$5 of the ticket is considered a donation.
- Consider setting up a web page for people to register, download forms, and learn more about the event. Also, Conquer Chiari can put all the information on the Conquer Chiari website.
- Think about different ways to raise money at the event. Keep ticket prices and sponsorship fees reason-

able, especially for the first year the event is held. To get the event established, it is better to have more participants and not worry as much about how much money is raised.

- Approach local businesses to sponsor individual holes (in exchange they get a sign at that hole); or businesses can sponsor prizes/handouts at holes. You could also have participants pay extra for hole-specific competitions. If allowed you can set up a food/beverage booth with donated baked goods, etc. Consider holding a 50/50 raffle as part of the fun and don't forget to ask straight out for donations.
- Consider having a secondary event after the golf, such as a dinner, cookout or silent auction. This works best if it can be held at or near the golf course.
- Start calling businesses for sponsorships!! This is where the work really starts. Use your contacts in the community to approach local businesses about supporting the event. Don't be shy about asking. If a business does not want to donate money, ask if there is a product they can donate for the prizes or goodie bags. Don't pay for anything out of pocket without asking a business if they will do it for free. Also, get comfortable asking outright for a donation. Practice your pitch so that you can clearly communicate the cause and the event. Recruit others to help with approaching businesses, it is one of the keys to a successful event. Make sure you acknowledge those businesses who helped in some way.
- Promote the event. Try to get the local paper and radio stations to promote the event. Also, Conquer Chiari can promote it within the Chiari community through our website and newsletter. Tell everyone you know about it and ask them to help spread the word. Promotion is one of the keys to a good event.
- Create a goodie bag for the participants. You can include whatever the local businesses donate, plus literature from the Conquer Chiari site. Make sure you have plenty of water on hand for the participants.
- Consider making up T-shirts for the walkers with the name of the event.
- The day of the event, arrive early to set up and make sure everything is organized.
- Try to collect all donations at the event itself. Checks should be made payable to 'C&S Patient Education Foundation' or 'Conquer Chiari'. Note, Conquer Chiari will send out receipts and thank-you letters to everyone who writes a check, but we can not issue a receipt for a cash donation. Make sure someone is appointed to handle the money during the event.
- Have fun at the event! Share stories with the participants and make sure everyone has what they need.
- After the event, try to get some feedback from the participants about how to improve the event for next year. Send out thank-you letters to the businesses, large donors and walkers.
- Start planning for next year! Events like this can grow over the years, so what may start as a small fundraiser can end up being a major event in the future.

Conquer Chiari will help in any way we can, just email director@conquerchiari.org or call 724-940-0116.

In The News : Ohio Advocate Gets Local Press

[Editor's Note: The following is reprinted with permission from the Dayton Daily News. The original article can be found at: [Dayton Daily News](#). Also, Carrie wanted everyone to know that she did not organize this event by herself and that Merideth deserves a lot of credit.]

By Kevin Lamb

Sunday, September 23, 2007

URBANA — The obscure disorder on Carrie Marenberg's new Web site afflicts about 300,000 Americans. That makes it more prevalent than cervical cancer, the target of ubiquitous TV advertising.

"And so many people more than likely have Chiari malformation, but don't know it," she said. Her own doctor only learned about Chiari from her MRI report last February.

Marenberg had to look it up on the Internet to find out it meant the bottom of her brain extended out of her skull, crowding the spinal cord and blocking the flow of spinal fluid to and from the brain. "Oh my gosh!" she said. "My brain is not where it should be."

After that first reaction, Marenberg made it her mission to help doctors and patients recognize Chiari.

- Her Web site offers information about the disorder and enables people to discuss their often-debilitating symptoms and contact other sufferers at www.freewebs.com/carrieschiariconnection/.

- She organized Chiari Awareness Day on Sept. 29 in Cincinnati, bringing patients together with expert speakers. The \$10 fee will raise funds for patients who need money for treatment and drugs, but often can't work.

"It's not only the general public that doesn't understand Chiari, but also the doctors," said Marenberg, who just turned 36 and works full-time building airplane lights at Honeywell International in Urbana. Her ultimate goal is for doctors to know enough about it to reflexively order diagnostic MRIs when patients have symptoms.

Until then, she said patients will need enough information to tell doctors, "Hey, what about Chiari?"

Patients typically endure decades of repeated misdiagnoses. Marenberg's headaches began as a toddler, but doctors ruled out migraines or sinus

problems. They've repeatedly scanned her brain, tested her blood sugar, checked her eyes and wired up her heart.

The symptoms worsened in the last four years, with dizzy spells, frequent fatigue and heart palpitations with slow heartbeats, skipped beats and racing ones "like when someone scares you and you gasp for air." She'd pass out, sometimes losing sight and hearing while still conscious.

Her concentration evaporated in a "Chiari fog," leaving sentences unfinished or getting lost in familiar areas. Her three teenagers would point out that she just said, "Go let the leash outside," when she meant the dog.

Her neck hurt constantly. It took four doctor visits for neck pain before the MRI needed to diagnose Chiari.

Marenberg could undergo surgery, removing skull and spinal bone to relieve pressure where her brain doesn't belong. But it's risky and doesn't always help, so she's trying drugs and procedures.

The worst pain comes suddenly, Marenberg said, and feels like her head "is about to explode. You grab your head, the top and sides, and push like you're trying to hold everything together."

Even without much symptom relief, knowing what's wrong helps. She knows not to ride any more roller coasters. She has learned to avoid the worst of that upper-neck pain by yelling less loudly at her son's football game. And she no longer doubts her sanity.

"It's very frustrating, hurting day after day and not knowing why," Marenberg said. "I felt like a hypochondriac. I also went into a very severe depression.

"So when I finally found out what it was, yes it was hard to swallow that I had a brain malformation, but at least I know what was causing me to feel the way I felt."

Glossary: Common Chiari Terms

central canal - very center of the spinal cord, so named because it starts as a hollow tube which closes in most people as they age

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

cervical - the upper part of the spine; the neck area

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

Chiari II - more severe form of malformation which involves descent of parts of the brainstem and is usually associated with Spina Bifida

cine MRI - type of MRI which can measure CSF flow

compliance - a measure of how much a vessel changes in volume due to a change in pressure; dV/dP ; the inverse of elastance

cranial nerve - one of 12 pairs of nerves that originate in the brain as opposed to the spinal cord

craniocervical junction - the area where the skull and spine meet

cranium - the skull

craniectomy - surgical technique where part of the skull is removed

decompression surgery - general term used for any of several surgical techniques employed to create more space around a Chiari malformation and to relieve compression

dura - tough, outer covering of the brain and spinal cord

dural scoring - surgical technique where a series of cuts are made into the dura, but the dura is not completely opened

duraplasty - surgical technique where the dura is opened and expanded by sewing a patch into it

graft - material, or tissue, surgically implanted into a body part to replace or repair a defect

hydrocephalus - a condition where there is an unusually large amount of CSF in the brain, resulting in swollen ventricles

ICP - intracranial pressure; pressure of the CSF inside the skull

intradural exploration - general term referred to a surgeon finding and removing any scarring or obstructions to CSF flow that exist underneath the dura

laminectomy - surgical technique where part of a vertebra is removed

lumbar - the lower part of the spine

magnetic resonance imaging (MRI) - diagnostic device which uses a strong magnetic field to create images of the body's internal parts

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

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

syrinx - fluid filled cyst in the spinal cord

thoracic - relating to the middle part of the spine, or chest area

tonsillar herniation - descent of the cerebellar tonsils into the spinal area; often measure in mm

ventricle - a CSF filled space in the brain

vertebra - one of the individual bones of the spinal column



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The C&S Patient Education Foundation is a 501(c)(3) non-profit organization dedicated to improving the experiences and outcomes of Chiari and syringomyelia patients through education, awareness and research. Our goal, quite simply, is to Conquer Chiari.

To support Chiari & Syringomyelia News, and the Conquer Chiari effort, you can make a tax-deductible donation at www.conquerchiari.org. Or send a check, made payable to C&S Patient Education Foundation to:

C&S Patient Education Foundation
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