

ASAP CONNECTIONS

Providing Research, Education and Support since 1988

Dilated Central Canal

With the advancement of MRI technology, individuals are being diagnosed with very small syrinxes that they were unable to detect in years prior. Some people will be told they have a slit-like syrinx or dilated central canal. These are generally 1-2 mm wide in diameter and are often described as a “nonexpansile” syrinx. To understand why this small cavity is present, a review of normal anatomy is helpful.

The spinal cord forms as a tube. On the inside of the tube is a small fluid cavity called the central canal. This disappears during development or during infancy. When the central canal does not completely close, a small leftover of the canal can be seen on MRI and is referred to as a ‘persistent central canal’. A radiology report may contain the phrase ‘a small syrinx versus a persistent central canal’. A repeat MRI may be recommended in order to determine a definite diagnosis. This often causes stress for the person waiting and hoping for a concrete diagnosis.

The current understanding leads physicians to recommend that if the dilation of the central canal is not associated with any other known cause of a syrinx (such as the Chiari malformation, a tethered spinal cord, or tumor) it is likely to be a benign finding that will not cause any symptoms and is not likely to grow or enlarge. To make sure, a follow up MRI is usually ordered. In cases where an underlying condition known to be associated with syrinx formation is present (such as a Chiari malformation) or if repeated imaging shows a change in the dilation, then it is more likely to be a true syrinx that should be evaluated and monitored accordingly.

*“I fight for my health every day
in ways that most people don’t
understand. I’m not lazy. I’m a
warrior!” – Unknown*

Cognitive Behavioral Therapy

Cognitive behavioral therapy (CBT) is a form of talk therapy that helps people identify and develop skills to change negative thoughts and behaviors. It is believed by changing the way we think and respond, people can change their awareness of pain and develop better coping skills, even if the actual level of pain stays the same.

CBT changes the way people view their pain. You recognize that the pain interferes less with your quality of life, and therefore you can function better. It can also change the physical response in the brain that makes pain worse. Pain causes stress, and stress affects pain control chemicals in the brain, such as norepinephrine and serotonin. CBT reduces the arousal that impacts these chemicals. This, in effect, may make the body’s natural pain relief response more powerful.

To treat chronic pain, CBT is often used together with other methods of pain management. These remedies may include medications, physical therapy, weight loss, massage, or in extreme cases, surgery.

If you want to try CBT for pain management, first talk with your doctor. He or she may know of a cognitive behavioral therapist who specializes in chronic pain or be able to point you in the right direction.

When you begin, the therapist will evaluate your pain, including the history and your current pain management methods. Your doctor will also do a general psychological assessment to identify any issues that may be making the pain worse.

Finding a CBT Therapist

Cognitive behavioral therapy is an increasingly popular treatment for all kinds of problems, pain relief included. As a result, more and more professionals call themselves cognitive behavioral therapists these days, even when they don’t have the proper training. To find a legitimate therapist who can help with pain management, but be sure to check credentials, conduct an interview and choose someone you like.

www.ASAP.org

KIDS For A Cure Club



Through a generous contribution and the efforts of ASAP member Annette Johnson, We have received a donation of children's books and plush toys. The items are being mailed to children with Chiari or syringomyelia having surgery or undergoing a hospital stay.

If you would like to nominate a child to receive an ASAP care package please email info@ASAP.org. Or you can send the name of the child, mailing address and date of hospitalization to the ASAP office.

Awareness Bookmarks

Get your free awareness bookmarks to share with other. Each contains a simple explanation of the disorders. Contact the ASAP office to order now!

Please Note: Articles in this newsletter are not intended as a substitute for medical advice and do not necessarily represent the viewpoints of the editor, Medical Advisory Board or Board of Directors. Please contact your doctor before engaging in any new therapy or medication.

Fundraising 2015

Molly's 2015 ASAP Walk and Roll for a Cure was held on Saturday, September 19, at the Bretton Woods Recreation Center in Germantown, Maryland. The event was a tremendous success. Molly Lichtenstein, who organized and coordinated the event, with support from ASAP President Patrice Schaublin and former board member John Caemmerer, was extremely pleased with the results.



"We had perfect weather, a beautiful venue, and a great turnout," Molly said. "I could not have been happier with the way things turned out." Patrice started things off with some brief introductory comments to explain the history of the ASAP organization, as well as providing the audience with some facts about Chiari malformation and syringomyelia. John followed as to why this cause is so important to him. Molly shared her story about what it is like to live with these conditions, and then it was time to walk. Once everyone had completed the walk, a raffle and a silent auction were held in order to raise more money for research and awareness.

Between donations, walk registrations, corporate sponsorships, and proceeds from the raffle and silent auction, the event raised more than \$21,000, as well as increased awareness of these rare conditions. "As anyone who suffers from a rare disease knows all too well, one of the biggest challenges in getting treatment and improving the way you feel is finding doctors who know anything about the condition," Molly said. "I am very proud and excited to be able to contribute to the research efforts by raising money and organizing events like this one to increase awareness. I am also extremely grateful to all the people who have been so generous, compassionate, and supportive by donating money and volunteering their time to help with the walk. Now, it's time for a much-needed rest, and then, in early 2016, it will be time to start planning for next year's walk!"





A Word From Our President

It's hard to believe another year is winding down. I know that's such a cliché, but it's true. I recall when I first made my phone call to ASAP as a terrified 32 year-old mother of two little girls and a newborn. Now, over 20 years later and I sit writing this column. I recently said to a woman that we have come a long way and she looked at me as if I was crazy. Looking back over the last 20+ years we have made great progress. We are getting the words Chiari and syringomyelia out there. It may not be as quick as we would like, but we are seeing it in print and on TV. More research is being done. More people are being diagnosed. Yes, we have a long way to go and with your help ASAP will continue to educate, provide support and fund research.

As we approach the holidays I want to first thank you again for the opportunity to serve as board president of our organization. I have met so many wonderful people. The holidays always make me think of the things for which we are grateful. I especially love Thanksgiving because it's a perfect time to be with family and friends with no expectations. As we approach December, it seems to get a tad more hectic. Between decorating, gift giving and holiday parties we can get lost in the meaning of what the holidays are all about.

We at ASAP are thankful for a wonderful medical advisory board, research committee, board of directors, staff and volunteers. I am so grateful for all your hard work. Many thanks to our members for the fundraisers they have done this past year and for those members and volunteers who have contributed their time and talents helping ASAP move forward.

I am amazed every year at the conference when I see the time our doctors take to sit with patients one by one and listen to each one's concerns. The lectures we hear are invaluable and the research we are doing is fantastic. This is thanks to all of the individuals who give so generously.

We have a lot in store for 2016. Our research involving the cavalier dogs and syringomyelia will continue with an update at our conference in July. We just funded a new grant with Georgia Institute of Technology, measuring CSF flow dynamics in real-time using a novel MRI sequence. This method allows us to look at the affect of respiration and cardiac rhythms to see how they regulate the CSF flow.

Thanks again to all for your continued support. On behalf of ASAP and myself, I would like to wish you all a blessed, happy and healthy holiday season.

Gentle Hugs always,
Patrice
Patrice_Schaublin@ASAP.org

Cooking Made Easier

For those who find cooking can bring on symptoms, try these helpful hints to make it less stressful on your body.

- Use a bar stool adjusted to the proper height so you can sit while chopping or stirring.
- Place ingredients like staples you use often on lower, easy to reach shelves.
- Use a toaster oven or microwave placed on the counter when possible to prevent bending to place and retrieve food from the oven.
- Hang pans and utensils you use most on wall hooks at arm level.
- Two-handled pots distribute weight across both hands to making carrying easier.
- Add a strip of padded tubing to handles to make grip more secure.
- Use an ergonomic knife with a large handle for cutting and chopping or a food processor.
- Instead of trying to carry a pot of water from the sink to the stove, place the empty pot on the stove and fill with a cup or small pitcher.
- Use a slow cooker for preparing meals. You won't have to stand over a hot stove stirring.
- When you cook, double the recipe and freeze the extra serving for another day.



If you would like to share coping techniques you have found helpful please send your suggestions to Patricia_Maxwell@ASAP.org.

***“DO NOT CONFUSE MY BAD DAYS AS A SIGN
OF WEAKNESS. THOSE ARE ACTUALLY THE
DAYS I'M FIGHTING MY HARDEST.”***

– UNKNOWN

Why is Chiari So Hard to Treat?

For many diagnosed with Chiari malformation (CM) the condition can be treated successfully. However there are others who even after surgery continue to have problems or develop new ones. One of the reasons for this could be that individuals can have other disorders, diseases or conditions that might affect the Chiari or be responsible for symptoms. They may go undiagnosed even after years of treatment, specialists and searching for answers.

Conditions sometimes linked with Chiari malformation are:

- **Aseptic meningitis** – Inflammation of the linings of the brain and spinal cord that is not caused by bacteria. It can occur after any surgery that requires the opening of the brain coverings. During surgery, some blood cells may enter the cerebral spinal fluid. After surgery, as these cells break down, the products of cellular breakdown irritate the inside lining of the meninges. This can cause a headache and fever and is treated with steroid therapy.
- **Basilar invagination** – This is when the upper end of the spine, or C2 vertebra, sticks into the skull putting pressure on the brain stem.
- **Connective tissue disorders** – Connective tissue supports many body parts, such as the skin, muscles and ligaments. When connective tissue is faulty, it is usually due to faulty collagen. Collagen is a protein that works like glue in the body. It makes connective tissue strong and stretchy. One of the connective tissue diseases that might cause CM is Ehlers-Danlos Syndrome (EDS). EDS is a genetic disease that makes joints too mobile, skin too stretchy and tissue too fragile. There are six different types of EDS categorized by the symptoms they cause. Treatment includes physical and occupational therapy to learn how to avoid injury.
- **Hydrocephalus** - The condition develops when the ventricles of the brain hold too much cerebrospinal fluid (CSF). Treatment includes placing a shunt to drain the fluid.
- **Intracranial hypertension** - Also known as pseudotumor cerebri, it is a condition characterized by CSF pressure in the brain always being too high. It can cause headache, nausea, vomiting and vision problems. The vision problems are due to optic nerve swelling. Optic nerve swelling can be found by an eye exam. A spinal tap measures pressure, but longer-term pressure monitoring may be needed. Surgery and a hospital stay are needed for pressure monitoring. Treatments include medical therapies to reduce CSF production or a shunt to drain CSF.
- **Myelomeningocele** - A birth defect that occurs when the vertebrae do not fully form and remain open allowing the spinal cord to protrude. The condition is also known as a type of spina bifida. Surgical repair is usually required.
- **Neuropathic pain syndrome** - Patients experiencing pain caused by damage to the central nervous system are said to have neuropathic pain syndrome. The symptoms are burning pain and abnormal feelings. Neuropathic pain syndrome is hard to treat, but responds best to medication that treats neuropathic pain.
- **Scoliosis** - Scoliosis, a side-to-side curving of the spine, is one of the most common first symptoms in pediatric Chiari patients over 3 years old. Early and quick treatment of CM in a child with fast progressing scoliosis can halt progress of the spine curving. These children also should have an MRI of the spine to assist in ruling out syringomyelia (SM), which may cause scoliosis.
- **Sleep apnea** - A common sleep disorder characterized by brief interruptions of breathing during sleep. These episodes usually last 10 seconds or more and occur repeatedly throughout the night.
- **Spina bifida** - When a person is born with a spinal canal that did not close during development, he or she is said to have spina bifida. There are many different types of spina bifida, ranging from spina bifida occulta, which is benign, to myelomeningocele, which is severe. Most patients with myelomeningocele will have Chiari II.
- **Syringobulbia** - A medical condition where syrinxes, or fluid filled cavities, affect the brainstem. This defect normally results from congenital abnormality, trauma or tumor growth. It mostly occurs within the lower brainstem as a slit-like gap. This may affect one or more cranial nerves, resulting in various kinds of facial palsies. Sensory and motor nerve pathways may be affected by interruption and/or compression of nerves. This disorder is associated with syringomyelia, a syrinx limited to the spinal cord.
- **Syringomyelia** - A disorder in which a cyst forms within the spinal cord. This cyst, called a syrinx, expands and elongates over time, destroying a portion of the spinal cord from its center and expanding outward. As a syrinx widens it compresses and injures nerve fibers that carry information from the brain to the extremities. Damage to the spinal cord often leads to progressive weakness in the arms and legs, stiffness in the back, shoulders, arms, or legs, and chronic, severe pain. Other symptoms may include headaches, a loss of the ability to feel extremes of hot or cold (especially in the hands), and loss of bladder and other functions. Each individual experiences a different combination of symptoms depending on where in the spinal cord the syrinx forms and how far it extends. Signs of the disorder tend to develop slowly, although sudden onset may occur with coughing or straining.
- **Tethered spinal cord syndrome** - This is a disorder in which the spinal cord is abnormally attached to a structure within the spine and is causing harmful traction on the spinal cord.

What to Expect after Surgery for Chiari with Syringomyelia

I see this question often on Facebook so thought it would be of interest to others. The following is a transcription from an ASAP teleconference presented by Dr. John Heiss, National Institutes of Health, Bethesda, Maryland. The study which was published in the Journal of Neurosurgery involved twenty-nine patients that were treated at the National Institutes of Health prior to 2009.

Background: The study was performed to find out how long it would take for a syrinx to become smaller after surgery. Patients enrolled into the study had syrinxes that were large enough to distend the spinal cord, with the average diameter of a syrinx being about 7 mm, which is about two-thirds of the diameter of a normal spinal cord.

A reduction of 50% in syrinx diameter was the primary outcome measure because this amount of reduction in syrinx diameter would indicate that the syrinx was no longer distending the spinal cord. They were also interested in what happens over time after surgery to major symptoms of syringomyelia such as weakness, numbness, and dysesthetic (neuropathic) pain.

All patients gave their informed consent to participate in the research study. The study was approved beforehand by the Institutional Review Board, which is a committee that evaluates protocols to see if the research is scientifically sound and that it does not expose patients to unnecessary risk.

For the research study patients with Chiari I and syringomyelia came to the Clinical Center of the NIH for their evaluation and treatment. Patients were 16 to 61 years of age. They underwent MRI scans of the neck before surgery; the diameter of their syrinxes was measured using a computer workstation. They also recorded a medical history and a neurological examination.

The presence and severity of signs and symptoms such as weakness, atrophy (wasting of muscles), spasticity (tightness of muscles), ataxia (unsteadiness while walking), pain and unpleasant sensations in the torso or extremities (dysesthetic pain), and loss of sensation was specifically noted.

MRI scan and a history and neurologic examination are standard studies for all patients who are being considered for surgical treatment of the Chiari I malformation and syringomyelia. Because of the way in which these tests were applied they could be used for research:

1. It was decided before the study began that all patients would have the same type of MRI study and their neurological examination recorded in the same manner.
2. The research was designed so information from the MRI scans and examinations was entered as each person was examined (prospectively) rather than from a retrospective analysis of charts on patients who had completed treatment.

3. Tests such as the MRI scan, history, and neurological examination were performed in the same way after surgery as before surgery and repeated at specified intervals (1 week, 3-6 months, 1 year, and yearly) after surgery.

The same surgical procedure, craniocervical decompression and duroplasty, was performed on all patients in the study. The average period of follow-up after surgery was 3 years. The number of MRI scans and examinations that were performed on each patient exceeded the number that would be performed on the basis of clinical care.

Six months after surgery all but one patient reported improvement. The patient who did not improve reported that he was stable. In all but 4 patients the syrinxes became less than one-half of their original diameters by 3-6 months after surgery. By 2 years after surgery all syrinxes had become less than one-half of their original diameters. The length of the syrinxes also became progressively shorter after surgery compared to before surgery.

Results showed that in twelve patients (41%), the syrinxes did not disappear completely. There was no difference in outcome in those without complete disappearance of their syrinxes compared to those with complete disappearance of their syrinxes. It was remarkable that a minority of patients became free of symptoms after surgery, only 22% at 3 months, 29% at 1 year, and 32% at 2 years after surgery. Residual signs and symptoms arose from residual dysfunction of the spinal cord. The most frequent signs and symptoms that persisted after surgical treatment were painful dysesthesias (neuropathic pain) and loss of sensation, which were found in about one-half of patients.

Conclusions from the study were:

1. Almost all patients notice some improvement after surgery.
2. Many patients experience residual symptoms, especially dysesthetic (neuropathic or chronic) pain and loss of sensation.
3. All syrinxes will become smaller over time after successful surgery, although many will not disappear completely.
4. Patients that have reduced-size but not complete disappearance of their syrinxes had similar clinical outcome compared to patients with complete disappearance of their syrinxes.

They do not believe that a small, collapsed syrinx will cause further injury to the spinal cord. Dr Heiss said they do not recommend additional surgery in patients whose syrinxes become smaller but do not resolve completely. The symptoms and signs reflect injury to the spinal cord that the syrinxes produced before surgery and not ongoing injury to the spinal cord.

Remember ASAP...

When It's Time to Remember Loved Ones

Our appreciation to everyone who made a recent donation to ASAP on behalf of their friends and loved ones.

We will send an acknowledgement card to individuals or families when you make a \$5 (or more) donation to the organization. Please indicate whether the gift is 'in honor of' or 'in memory of' and provide name and mailing address of the recipient.

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ASAP History

The 1998 ASAP conference was truly a celebration of ASAP's 10 years of growth. From the opening reception to the closing banquet there were many special touches that made the conference the best ever according to many attendees. It was also the largest attendance to date with approximately 170 people.

It was the dawn of social media and ASAP members communicating with each other through a listserv sponsored by the University of Maine faculty member Steve Godsoe. This was a chance for individuals who connected almost daily to meet face-to-face for the first time.

Attendees also received a copy of the anniversary book which contained a history of ASAP's first 10 years with photos and information compiled by Candace Morse.

Don White, Henry and Blanche Luckraft officially cut a special 10th anniversary cake decorated with the ASAP logo. The honor was awarded to them as the only individuals who had attended all ten conferences.

New Awareness and Fundraising Walks

by Patrice Schaublin

Going back to our roots, this past year has seen an effort to keep syringomyelia in the fore. We have made strides by getting state proclamations declaring May Syringomyelia Awareness Month. Additionally, ASAP started a campaign called Syringo-My-What?? Steps across the States. It is our hope that each state will eventually be represented in this walk.

We have also added another campaign, "Kicks and Licks," so I now need you to grab your sneakers and your leashes and join us for a walk/run/gathering at a park near you! Dogs are welcome! This is a national campaign. We will unite and start spreading awareness on the devastating diseases known as Chiari-malformation and syringomyelia. These diseases affects dogs, most commonly the King Charles Cavalier Spaniel, as well as their human counterparts. It also affects over 15 others breeds. The dogs have a Chiari-like malformation and syringomyelia. Dr. Lauren R. Talarico DVM Diplomate, ACVIM-Neurology from VCA SouthPaws Veterinary Specialists and Dr. Robert Keating, Children's National Medical Center will be working together with ASAP to help determine the underlying cause for this disease. Ultimately, this research will help further treatments that could potentially impact humans as well as man's best friend.

For those interested in hosting their own walk, please contact patrice_schaublin @ASAP.org.

Great times ahead for ASAP and our 4-legged friends. We will continue to work together in search of a cure!

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“Yes, I want to make a difference!”

I support the work of the American Syringomyelia and Chiari Alliance Project, Inc.

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Online Resources for Students with Disabilities

With more adaptive technologies and progressive legislature, prospective college students with disabilities have countless resources available to make the transition to higher education less stressful. Affordable Colleges Online just updated their College Resources for Students with Disabilities guidebook. You can see the guide here:

<http://www.affordablecollegesonline.org/college-resource-center/resources-for-students-with-disabilities/>

They've also created a complementary guide to scholarships and financial aid for students with disabilities to help students and their families better understand the vast number of financial aid options available to them. Key elements of the guide include:

- A comprehensive review of all of the scholarships available, listed by disability
- Amounts awarded and deadlines for each scholarship
- Debt-forgiveness options for those who acquired a disability post-graduation

You can find this guidebook at:

<http://www.affordablecollegesonline.org/college-resource-center/affordable-colleges-for-students-with-disabilities/>

[The links are also available at www.ASAP.org under Resources.]

Go Green

Tired of waiting for the newsletter to be delivered to your home address? Receive it up to two months faster. Go to www.ASAP.org, on the home page click on **Support and Resources** (the second box positioned above the pictures). That will take you to a landing page with a blue box on the right side. This is a list of connecting pages. Scroll down until you find **Go Green**. When you click **Go Green** it will take you to a page with information about **E-Connections**

(which is simply a PDF of the **ASAP Connections** that is mailed to you). The advantage of downloading the PDF file is you see the newsletter in color.

After you fill in the required information, hit submit and we will change your newsletter preference. Or you can just call 1-800-272-7282 and leave a message. Give your name, email address, zip code and say go green. Please repeat your name and email address to ensure we get the information correctly.



ASAP's Mission: to improve the lives of persons affected by syringomyelia, Chiari malformation and related disorders while we find the cure.

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ASAP Connections

The American Syringomyelia & Chiari Alliance Project (ASAP) is a tax-exempt 501(c)(3) organization. Our goals include providing a clearinghouse for information on syringomyelia (SM), Chiari malformation (CM), and related conditions.

We offer a supportive network of programs and services and fund research to find better therapies and cures. ASAP is supported by tax deductible donations.

ASAP Connections is published quarterly for ASAP members. Your contributions of articles, letters, and photos are encouraged. The editor reserves the right to edit any article in order to accommodate space.

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