

ASAP Funds New Research

ASAP Board Grants Three New Research Projects for 2020

Quality Indicators in Pediatric Chiari I

Principal Investigator: Gerald Grant, MD
Institution: Stanford University Medical Center
Grant: \$102,568

Lay Summary of Research:

Chiari I malformation is an anatomical abnormality characterized by a number of anatomic and radiologic findings. Classically, children can present with headache, long tract signs (such as loss of sensation, position sense, weakness, spasticity, or incontinence), bulbar symptoms (such as difficulty swallowing, dysarthria, or other dysfunction of the lower cranial nerves), or other neurologic impairments. There are also impairments across physical, psychological, and social domains in patients with Chiari I.

Surgical decompression is a common neurosurgical procedure performed to alleviate Chiari symptoms, however many controversies exist over the implementation of treatment. Our goal is to establish the “standard of care” for the surgical management of Chiari I malformation. Despite quality of life measures to assess the surgical outcomes of Chiari decompression, the natural history is relatively unknown. We will use an evidence-based approach to establish Chiari indicators using a Collaborative Health Outcomes Information Registry to inform clinical and surgical decision-making.

We have reviewed the literature in an unbiased and systematic fashion to identify variables that can either predict whether a patient will benefit from surgery or whether a surgery has been “successful.” We will apply these variables prospectively before and after surgery.



Quantifying Gait and Postural Control in Chiari Patients

Principal Investigator: Brian Davis, PhD
Institution: Akron Children’s Hospital
Grant: \$49,896

Lay Summary of Research:

Chiari malformation (CM) is a disorder at the junction of the skull and spine. It is associated with protrusion of the base of the brain into the top of the spine, thus inhibiting flow of cerebrospinal fluid. This results in pressure buildup, causing headaches, dizziness, difficulty swallowing, muscle weakness, and loss of neuromuscular coordination. The last of these symptoms is the focus of the proposed research.

Specifically, this study aims to quantify postural stability in pediatric patients with CM. Although loss of stability and impaired walking are frequent symptoms, no previous studies have examined either of these deficits in CM patients. Understanding neuromuscular control will provide further insights to neurologists, neurosurgeons planning surgical intervention and to therapists designing pain reduction and restorative balance techniques. Between 2012 and 2019, 192 patients were treated at Akron Children’s Hospital (ACH) with a diagnosis of Chiari syndrome without spina bifida or hydrocephalus.

The proposed project will recruit 20 of these pediatric patients with SM and 20 age and sex matched controls and measure neuromuscular coordination using two simple tests including (i) the ability to stand upright and motionless for 30 second durations and (ii) the ability to perform simple walking trials across an instrumented walkway. Data will be collected at baseline and again at six months to document (i) differences in age-matched control subjects and (ii) changes over time. This data will serve as a baseline for all future studies that examine neuromuscular coordination in CM patients and help inform clinical practice using objective measures of symptoms and their management.

2020 Research

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The team that has been assembled for this research project includes Dr Gayathri Sreedher, an experience pediatric neuro-radiologist based at ACH (the 7th largest pediatric hospital in the USA), Dr Brian Davis, former president of the International Society of Biomechanics and co-author of a textbook on human gait, and Dr Doug Wajda from Cleveland State University and an expert on fall risk assessment.

Amplified Imaging of the Pediatric Brain

Principal Investigator: Michael Moseley, PhD
Institute: Stanford University, Department of Radiology
Grant: \$90,728

Lay Summary of Research:

The research project will create and test a new way of illustrating and mapping small brain dynamics using a novel MRI method, “amplified Magnetic Resonance Imaging” (aMRI). This method takes conventional series of MR images of the pediatric brain and processes these into a ‘cine’ or movie loop that can amplify intrinsic motions from the pulsing arteries on the surrounding brain to detect a variety of subtle brain and spinal cord dynamic abnormalities. We anticipate that the applications of amplified movies of the brain will become an important tool in pediatric imaging and offer unique visualizations of altered or abnormal structure and function in potentially any tissue from any modality. We expect that this new tissue diagnostic mechanism will rapidly advance precision health in children.

**ONE POSITIVE THOUGHT
IN THE MORNING
CAN CHANGE THE WHOLE DAY.**

Stamp Project

ASAP members and friends have been collecting postage stamps as a fundraiser since the early 90s. It is an easy and fun way to make a difference. Stamps are sold to collectors and stamp clubs with the proceeds going to support programs for the Chiari and Syringomyelia community.

Please send cancelled postage stamps that are in good condition (attached to envelope and trimmed with a 1/4 to 1/2 inch border around undamaged stamp). Forever, special occasion and foreign stamps are accepted as well as postcards. Nonprofit, postage meter and presort stamps are not collectable.

**Mail stamps to:
Charles Petkevich
ASAP Stamp Project
6202 SW 2nd Court
Plantation, FL 33317**



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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.

www.ASAP.org

Visit us on the web and be sure to check out our YouTube and FaceBook groups.



A Word From Our President

Eric J Berning

Hello everyone,

Let me start by saying a BIG THANK YOU to everyone who produced, spoke, attended, and in so many other ways helped support our virtual conference this year! All told, we had over 500 individual participants from across the globe for our 32nd annual ASAP conference. What an enormous success in advancing our mission of education, research, and support for the CM/SM community. We are still compiling the Q&A session questions that we were unable to get to in real time so that we can post them for you, but the conference videos are now up and available for viewing and sharing. Be sure to mark your calendars for next year's conference.

July 21-25, 2021
hosted by Dr Konstantin Slavin
at Chicago's historic Knickerbocker Hotel

We will be back "live and in person" and encourage everyone to join us. We are looking into the possibility of a virtual offering as well, but have not ironed out the details yet—so stay tuned.

We are now heading into my favorite time of year: FALL.

September is Chiari Awareness Month, when we typically hold most of our fundraising walks to raise awareness and advance the important mission of our organization. This year, due to current circumstances, we have two "virtual walks" taking place: Molly's walk in Maryland and Jeni's walk in Florida. Please visit the ASAP website for links to both and let's show our support for each of them.

I also love fall because of the change in the temperature. Here in Houston we go from oppressively hot to "slightly less hot" as we prepare for the one weekend in late December/early January when winter shows up for a brief visit. All kidding aside, the drop in the heat level is welcomed as it is much more comfortable for me to function—I'm sure many of you can relate. As I write this my son is about to start his senior year of high school, and I am looking forward to cheering on all his activities until we have to get out the tissues for his graduation. I'm not crying—that's just pumpkin spice in my eye.

Did I mention football season? (Go Browns!)

Of course, even with some things returning to a modified version of "normal," we are facing a different environment than we're used to. The COVID-19 pandemic and life in it has me thinking about one of my favorite movies/books of all time, *James and the Giant*

Peach. One line from the movie has always stood out to me. James is lying on the beach, gazing at the clouds with his father. His dad asks if he can see the Empire State Building, but James does not. "Well then," his dad suggests, "look at it a different way." James can see it. So how does this scene tie in to the pandemic? Let me explain: the current lockdowns are, in many ways, similar to what we with "invisible illnesses" experience every day.

Trying to explain to others what life is like with an invisible condition can be extremely difficult. On one hand, you don't want to sound like a complainer, but on the other you may hear the common response, "Well, you don't look sick." But, looking at it a different way, maybe the lockdowns can be used as an educational tool to help us explain what we experience daily. "Stuck at home, unable to do anything" is what we experience when our pain limits our ability to do what we would like. While staying home from work or school and being in front of the TV may sound like fun, when it's your only option it gets old quickly.

Our apprehension of doing something "normal" that may result in a debilitating trigger of our symptoms causes us to "mask" our willingness to participate so we can avoid any issues. Those who do not deal with our conditions but who are now taking extra precautions to safeguard themselves (masks, social distancing, etc.) may be able to understand better the challenges we face on a day to day basis to prevent harm to ourselves.

Let's look at this "a different way" and use this opportunity to help others "see it." Because of our unique perspective, we can also help lift people up by reassuring them that the current circumstances DO NOT DEFINE US and that "while you may have to live within limits, you can still lead a limitless life!" Virus or no virus, we all have control over only one thing in life: our response to our circumstances. Everything in life has external influences; we can only control how we react to them. Let's react positively and help others to do the same. After all, we're in this together.

One final note: ASAP is looking to expand our Board of Directors and welcome all who are interested to submit an application for consideration. Please email me at eric_berning@ASAP.org and we will get you the information you need.

Now go live that limitless life!

ASAP Conference 2020

With over 700 registered to take part in the 3-day event in July, our annual conference went virtual.

For the past 31 years the ASAP conference has brought together the world's foremost authorities to present their research, experiences and knowledge about Chiari, Syringomyelia and related disorders. This year we were excited to be able to offer this special opportunity to people all over the world. Twenty-four speakers shared their knowledge, volunteered their time and made the event a huge success.

We knew that holding the meeting virtually, we would miss the personal contact that is so much a part of our conference each year. So it was important to select a program that would allow face to face interactions while maintaining social distancing. For most of us this was a new experience. But technology allowed us to reconnect with friends and make new ones. It brought the experts into the homes of hundreds eager to learn more about these conditions that effect thousands.

The videos are now available on our YouTube channel and include five Question and Answer sessions presented during the conference.

www.youtube.com/channel/UCJHul2aTWLIjLBuUisJSWvg

We look forward to seeing you all in July 2021 in Chicago, Illinois at the Knickerbocker Hotel.

Comments from attendees:

Thank you so much, please keep plugging away I need this so much more than oxycodone and methadone for pain.
Richard R.

I enjoyed this virtual conference, ironically I'm in the hospital just had my shunt replaced yesterday. thank you ASAP and all the wonderful Drs who volunteer their time each year for ASAP's conference. I've never been able to attend but always wanted to. *Karen G.*

Thanks for great virtual conference. I missed the interactions in person, but enjoyed the format. *Wendi M.*

Thank you doctors and everyone. It was a great conference!
Heather W.

Very informative! Thank you to all who put your time/energy into this learning experience! God Bless Everyone.
Karen W.



2020 Conference Speakers:

Allison Ashley-Koch, PhD, Duke Institute for Brain Sciences

Ulrich Batzdorf, MD, UCLA School of Medicine

Ann Berger, MD, Pain and Palliative Care/ National Institutes of Health

Paolo Bolognese MD, Chiari Neurosurgical Center

Brian Dlouhy, MD, University of Iowa Children's Hospital

Gerry Grant MD, Stanford Children's Health and Lucile Packard Children's Hospital

Barth Green MD, University of Miami Medical Center

John D Heiss, MD, NINDS/National Institutes of Health

Bermans Iskandar, MD, University of Wisconsin, Madison

Robert Keating, MD, Children's National Medical Center

Petra Klinge, MD, Warren Alpert Medical School / Brown University

Dave Knowlton, CERI Cannabis Education & Research Institute

Roger Kula, MD, Chiari Neurosurgical Center

Jorge Lazereff, MD, Mattel Children's Hospital / UCLA

Michael Levy, MD, Rady Children's Hospital / San Diego

Cormac Maher, MD, University of Michigan, Ann Arbor

Anne Maitland, MD, Mount Sinai Hospital

Bryn Martin, PhD, University of Iowa

John Oró, MD, Chiari Care Institute, Aurora

Konstantin Slavin, MD, University of Illinois Chicago

Ian Stein, MD, Neurological Specialties of Long Island

Erol Veznedaroglu, MD, Drexel Neurosciences Institute

Thank you to all our speakers for their time and knowledge to assure we are all better prepared for tomorrow.

A Caregiver's Bill of Rights

I have the right

- **to take care of myself. This is not an act of selfishness. It will give me the capability of taking better care of my relative.**
- **to seek help from others even though my relative may object. I recognize the limits of my own endurance and strength.**
- **to maintain facets of my own life that do not include the person I care for, just as I would if he or she were healthy. I know that I do everything that I reasonably can for this person, and I have the right to do some things just for myself.**
- **to get angry, be depressed, and express other difficult feelings occasionally.**
- **to reject any attempt by my relative (either conscious or unconscious) to manipulate me through guilt, anger, or depression.**
- **to receive consideration, affection, forgiveness, and acceptance for what I do from my loved one for as long as I offer these qualities in return.**
- **to take pride in what I am accomplishing and to applaud the courage it has sometimes taken to meet the needs of my relative.**
- **to protect my individuality and my right to make a life for myself that will sustain me in the time when my relative no longer needs my full-time help.**
- **to expect and demand that as new strides are made in finding resources to aid physically and mentally impaired persons in our country, similar strides will be made toward aiding and supporting caregivers.**

More Comments FROM the 2020 Conference

Thank you for everything. It was fantastic! Perfect work, *Greeting from Anna in Warsaw*

Thank you so much to all of the organizers and presenters. Well done. *Jeni A.*

Thank you to all the doctors and ASAP board members for bringing this conference to us virtually, hope to see another virtual conference in future. *Vicki C.*

Thank you so much for the conference, it really makes a great difference. *Tamara B.*

Timothy M George Fellowship Award

The Board of Directors voted unanimously to rename the ASAP Chiari and Syringomyelia Fellowship in memory of Timothy M George. Dr George was well known in the Chiari /Syringomyelia community for his knowledge, dedication and care.

The award is meant to assist medical students and neurosurgeons in the clinical investigation or basic science research of Syringomyelia, Chiari malformation and related disorders. The American Syringomyelia & Chiari Alliance Project, Inc. sponsors this fellowship award in the amount of \$10-20,000.

The fellowship is open to all neurosurgical residents and fully trained neurosurgeons at any stage of their career in the United States. The fellow will spend 6 to 12 months under the direction of a specific sponsor in clinical, translational, or basic science investigation as relevant to syringomyelia, Chiari malformation and related disorders.

Those interested in applying for the award can download instructions and application from the ASAP website or contact the ASAP office at info@ASAP.org or by calling 903-236-7079.

ASAP Board Welcomes New Member

Brian J. Dlouhy, MD is a pediatric and adult neurosurgeon at the University of Iowa Hospitals & Clinics and Stead Family Children's Hospital in Iowa City, Iowa. A graduate of New York University School of Medicine in 2007, he completed his neurosurgery residency at the University of Iowa Hospitals and Clinics. After finishing his residency in 2013, he completed an endoscopic and minimally invasive fellowship in Sydney, Australia followed by a pediatric neurosurgery fellowship at St Louis Children's Hospital and Washington University in St. Louis, Missouri. He joined the Department of Neurosurgery at the University of Iowa in 2015 as an assistant professor.

Dr Dlouhy treats all neurosurgical disorders but specializes in the treatment of Chiari malformations, syringomyelia, and disorders of the craniovertebral junction in patients of all ages, from infants to adults. This has led to numerous manuscripts published in peer-review journals and book chapters on the these topics.

In addition to his clinical activities, Dr Dlouhy leads a human brain research lab to better understand Chiari malformations and cerebellar/brainstem function. A goal of his research is to better understand the causes and pathophysiology of Chiari malformations and determine better treatment strategies.

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 person you would like to honor.

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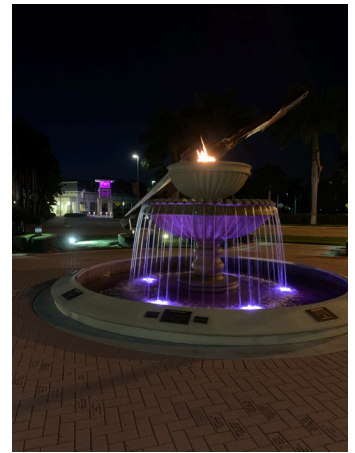
"Forget Me Nots"

Jeana Mackel

Robert Schloss

Ira Schloss

In Honor of
Chiari Awareness
Go Purple



Ask The Experts

We had so many great questions during the Q&A at the 2020 conference. However, due to time allotment we were unable to address them all. We will publish as many answers as possible on our website and in our newsletter.

Q: One of Dr. Maitland's slides noted anti-interleukin monoclonal antibodies for MCAS treatment, but didn't say which specific ones. Could she specify which meds are appropriate?

A: dupilumab - anti-IL4/13 (A. Maitland)

Q: What psychological therapies do you feel work best for pain? I hear a lot about mindfulness.

A: Mindfulness and cognitive behavioral therapy in combination can be very helpful.(A. Berger)

Q: Have you met in your studies a patient suffering from Chiari Syndrome and /or Syringomyelia who was treated with medical marijuana for chronic pain?

A: We have not, as yet, done any studies relating to patients with Chiari syndrome and medicinal marijuana. I believe as I mentioned during my talk, ASAP has asked us to consider undertaking such a study on their behalf to get some real data on this. That being said, there is at least one peer-reviewed study in this regard which showed a beneficial effect from medicinal marijuana for those suffering from Chiari syndrome. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5007175/> (D. Knowlton)

Q: Where and how can we find out if we have Celiac disease?

A: Blood tests are available for the diagnosis of Celiac. A gastroenterologist can perform an EGD (an esophagogastroduodenoscopy) to examine the lining of your esophagus, stomach, and duodenum and biopsy test results. (Dr. Maitland)

Q: Do edibles work as well for pain? I heard vaping is the best way.

A: I am told that edibles work as well for pain as "smoking a joint" with the caveat that the ingested route to having a physical impact is through the liver which slows it down. Folks who "take too much" cannabis when doing edibles usually do so because they do not receive an effect fast enough and take too much.

One further warning – there is pretty strong evidence that vaping has some harmful health effects, especially if the vape cartridge was produced with an oil base. There are vape cartridges that are flower based which have not been described with that problem. (D. Knowlton)

Q: Does depression increase your pain? Is there a medicine that treats both depression and pain?

A: Yes. Depression, sleep disturbances and pain are very linked. {For treatment I} would use tricyclic antidepressants, duloxetine or venlafaxine. (A. Berger)



Q: Have you ever heard of using a low energy shock waves for treatment of pain ? If so is it successful?

A: Yes. It can be useful. (A. Berger)

Q: What about taking medical marijuana/CBD & other medications since more than 90% of meds are metabolized by the cytochrome P450 complex. Should there be a time gap between taking these & any meds?

A: You are correct – Cannabidiol is one of the inhibitors of the cytochrome P450 modulator enzyme CYP2D6. You are now way beyond my pay grade! This is one of the reasons that I so strongly advocate that folks planning to take medicinal marijuana/CBD and combine them with other medications do so under the medical supervision of their doctor and often their pharmacist. (D. Knowlton)

Q: What psychological therapies do you feel work best for pain? I hear a lot about mindfulness.

A: I have found mindfulness and cognitive behavioral therapy in combination can be very helpful.(A. Berger)

Q: Are there differential diagnosis that mask or possibly are Mast Cell Activation?

A: yes- The MCA diagnosis is sometimes applied to patients with vague yet suggestive symptoms. These patients may suffer from an unrelated, overlooked disease. Certain cardiovascular disorders, endocrine disorders, neoplasms, GI diseases, primary skin diseases, infectious diseases, and neurologic or psychiatric disorders are among the numerous conditions sometimes confused with MCA. (A. Maitland)

Q: When gabapentin does not help the neuropathic pain what options are there?

A: Several other anticonvulsants, tricyclic antidepressants, duloxetine, baclofen, mexilitine, ketamine, clonidine, lidocaine patch, naltrexone, tramadol (A. Berger)

Thank you, Dr Anne Maitland, Dr Ann Berger and Dave Knowlton, for answers these questions.



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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