The Ataxia Center recently celebrated its 10th year of providing patients with ataxia and their families’ care and support in a multidisciplinary environment. A research symposium for physicians, researchers and other healthcare team members took place on October 15th, which was followed by a keynote address and reception attended by patients, their families and friends, and the Ataxia Center team. The goal of the symposium was to foster greater collaboration among the experts at Johns Hopkins University who research and treat neurodegenerative diseases. The Center is optimistic that this sharing of information will advance all of our research efforts to improve the diagnosis and management of ataxia. The research symposium also offered all in attendance the opportunity to hear from several experts during four sessions that included presentations on the translation of basic science research to patient care; rehabilitation-based interventions in ataxia, clinical innovations in ataxia, and research at the Johns Hopkins Ataxia Center.

Liana S. Rosenthal, M.D., the Director of the Ataxia Center, was recognized for innovative approaches in the treatment of patients with ataxia, as well as her management and development of the Johns Hopkins Ataxia Center. “Now that we have looked at the past 10 years of our work, we look towards the next 10 years to continue to improve our clinical care and research,” Dr. Rosenthal noted.

Donald Dawn, of the Gordon and Marilyn Macklin Foundation, introduced David Zee, M.D., of the Johns Hopkins Ataxia Center, who delivered the keynote address titled, “Why One Should Choose Ataxia as a Neurological Subspecialty.” Dr. Zee encouraged neurology residents to pursue the rewarding work focusing on ataxia as their career just as he did 50 years ago. He also thanked Mr. Dawn and the Gordon and Marilyn Macklin Foundation for their philanthropic commitment which provides funding for the Ataxia Center’s work.

Following the symposium, patients and guests were welcomed to a reception where over 100 people gathered to meet and greet and discuss ataxia research and treatment with researchers, neurologists and clinicians.

For more information on the Johns Hopkins Ataxia Center, please visit the webpage at: https://www.hopkinsmedicine.org/neurology_neurosurgery/centers_clinics/movement_disorders/ataxia/index.html
Gluten-free diets have become more popular over the last decade. Many people, including celebrities and the authors of bestselling diet books, have touted the benefits of gluten elimination for numerous symptoms. Since 2011, the availability of gluten-free food products has been steadily increasing, making this diet easier to follow. Patients with ataxia often ask whether a gluten-free diet might be helpful for their symptoms.

Diagnosis
All patients with ataxia from an unknown cause should have bloodwork done to look for treatable causes of their symptoms. Testing for gluten-related antibodies should be part of this workup, particularly in people with gastrointestinal symptoms, a history of autoimmune disease, or an immediate family member with celiac disease.

Should I Go Gluten-Free?
If antibodies are found in your blood and you are diagnosed with celiac disease or gluten ataxia, the answer is almost always YES. Following a strict gluten-free diet helps prevent damage to the intestine and may significantly improve ataxia symptoms. Studies suggest that it may take up to one year on the diet for improvements to be noticed. Consultation with your neurologist and a nutritionist is recommended.

If you have a genetic ataxia (i.e. Spinocerebellar Ataxia), there is no evidence that a gluten-free diet will help with your symptoms. Given that good nutrition and avoiding weight loss is particularly important in ataxia, attempting to follow a gluten-free diet may be difficult or even harmful for some patients, and is generally not recommended for genetic ataxias.

Definitions

Gluten: A protein found in wheat, barley, and rye. Foods rich in gluten include bread, baked goods, pasta, cereal, and beer. Gluten can also be found in unexpected products, including soup, salad dressing, food coloring, and even medications.

Celiac Disease: An autoimmune disorder in which the body triggers an immune response to gluten, causing damage to the small intestine. This results in poor absorption of nutrients and can lead to numerous health consequences. Gastrointestinal symptoms can range from mild to severe. Ataxia may also be a symptom in 10-12% of patients. Celiac disease is diagnosed by testing for certain antibodies in the blood, and conducting an intestinal biopsy to confirm. There is a genetic component to celiac disease, and genetic testing may be useful in some cases. Patients with celiac's must follow a very strict gluten-free diet for life.

Gluten Ataxia: Ataxia which occurs as a symptom related to gluten sensitivity is separate from celiac disease. Symptoms in gluten ataxia start very gradually and typically begin in a person's 50s. There may or may not be gastrointestinal symptoms along with ataxia, and not all patients have signs of intestinal disease on biopsy (unlike celiac disease). An antibody called “antigliadin” is detectable in the blood. Most patients with gluten ataxia will stabilize or improve with a strict gluten-free diet.
Ataxia Center News

Jana Arreola, CMA

We are pleased to introduce Jana Arreola to the ataxia community as the new Ataxia Clinic Coordinator for the Johns Hopkins Ataxia Center. Jana has been with Johns Hopkins University’s Movement Disorders Division serving patients since 2015. She is a certified medical assistant and has several years of experience as a medical office coordinator. For the past few years, she has been serving as the ataxia clinic’s “behind the scenes” assistant so some of you may have met or spoke with her previously.

Jana has coordinated the patient appointments and schedules of several neurologists in the Movement Disorder Division, and is currently, in addition to her role as the Ataxia Center’s Clinic Coordinator, the medical office coordinator for Dr. Emile Moukheiber, who is a neurologist at the Ataxia Center.

Jana’s desire to help patients and their families continues to develop, as well as her desire to work in the Ataxia center. In her spare time, she enjoys spending time outdoors with her children, family and pets, including a rabbit and her hound-mix dog named Armor.

Katherine Iannuzzelli

We are pleased to welcome Katherine Iannuzzelli as the Ataxia Center’s research assistant. While she previously worked with Dr. Rosenthal on Parkinson’s disease research in 2016, she is very excited to transition into Ataxia research. She will be working on several Ataxia-related research projects, including Dr. Marvel’s research on cognition and emotional regulation in Ataxia. Katherine graduated in May 2018 from Georgetown University with her Bachelors of Science in International Health. At Georgetown, she worked as an Emergency Medical Technician and completed a semester-long research practicum in rural Western Australia. She is applying to medical school next year and hopes to incorporate everything she has learned from the Ataxia Center’s multidisciplinary approach in her future as a physician. Outside of work, Katherine practices yoga and enjoys spending time with her friends and family.

Gerson Suarez-Cedeno, MD

Dr. Suarez-Cedeno is a movement disorder fellow and graduate of the Universidad de Antioquia Medical School of Medellin, Colombia. While there, he performed research of neuroprotective effects of antioxidants against toxins that cause Parkinson-like diseases. After graduating, he worked as a primary care physician and community educator for the Colombian Red Cross in the Amazon region. Then in 2014, he moved to the United States for his postgraduate neurology residency at the University of Texas Health Science Center at Houston. Dr. Suarez-Cedeno is interested in Parkinsonian syndromes, Ataxia, Deep Brain Stimulation and healthcare disparities. Outside of neurology, Dr. Suarez-Cedeno is a runner and salsa dancer.
How is cerebellum involved in cognition?

When we learn to move our body parts as a baby or as a child, a “map” of the world and the movements our bodies make are created in our brain. The cerebellum helps to correct errors when we make a mistake in a movement, for example, take a longer step than necessary when we walk, or spill water from a cup by holding it the wrong way and so on. Scientists believe that cerebellum does this same “error-correction” for our thinking and behavior as well. Thereby, it tempers down overt gestures, inappropriate and uninhibited behaviors. In cerebellar disease, this fine-tuning is lost. An erratic response of thought and behavior similar to the overshooting or undershooting of limb movements as seen in ataxia.

Cerebellar Cognitive Affective Syndrome (CCS)

Cerebellar cognitive affective syndrome (CCS) or Schmahmann syndrome, after the proposer of the idea Dr. Jeremy Schmahmann, includes a constellation of symptoms particularly seen in strokes or inflammation of cerebellum. The features of the syndrome can be grouped under four categories: executive, spatial, language and personality changes. 

Executive dysfunction makes it difficult to plan steps of an action, organize a thought, and adapt to changes in surroundings. Dwelling for too long on the same idea or conversation with pointless repetition can also be seen. 

Visuospatial problems make it difficult to navigate complex spaces (e.g., aisles of a supermarket, city roads, and freeway exit/merging). It may also impair her ability to draw, or find keys or similar things from among a heap of other items on the table. Language deficits can manifest as problems in grammar, abnormal sentence structures and loss of emotional content of speech. Personality changes in cerebellar diseases that could be visible includes a flattening of emotions or disinhibited behaviors such as overfamiliarity with strangers. We also see an increase in irritability and sudden outbursts that can cause problems in relationships. It should be noted that not every person with ataxia will demonstrate all of these features. More commonly we see a mix-and-match of varying degrees of two or three features from the above list.

Spinocerebellar Ataxias (SCAs)

Spinocerebellar ataxias (SCAs) affect the cerebellum in varying degrees, often causing it to shrink and lose its connection to the higher part of the brain. It should be noted that not all SCA show the same cognitive and mood-related symptoms. For example, SCA 6 and 8 show mild executive dysfunction but do not cause major memory problems. Whereas SCA 1, 2, 3, and 7 are known to cause more extensive problems in memory and behavior especially as the patients grow older with the disease. SCA 12, and 17 fall in the extreme end of the spectrum where dementia is a major feature of the syndrome.
When should you consider treating mood disorder in Ataxia?

The answer is, whenever the patient or the caregiver finds that it has become disruptive to normal life. Trained specialists can pick up many signs and symptoms in the clinic, but it does not mean all of them need to be treated. Your doctor may also order tests to look for other causes and as precaution before starting drugs. Though there are no FDA-approved medications for ataxia per se, animal studies have shown that certain mood-medications could improve the function of cerebellum and have been used off-label in ataxia - for instance, if an ataxia patient has anxiety or depression, an anti-depressant may help both balance and mood. In some instances, “fear of falling” may be so severe that the patient may not be inclined to participate in balance-physical therapy. Treating the anxiety in such cases may be considered.

What can be done about cognitive changes in ataxia?

There are no drugs or interventions available yet to replace lost or damaged brain cells. However, certain medications used in Alzheimer’s and similar dementias may be used to improve attention and concentration. This can improve memory in the short term. Building a cognitive “reserve” by being intellectually active is one way of resisting dementia. Cognitive therapy (usually performed by speech therapists) can help learn memory tricks, and employ shortcuts in remembering things of daily life. Discussions around driving can be tricky but need to be addressed early on to prevent accidents. When attention and ability to adapt to changes are affected, the brain makes use of “automatic systems”, learned behaviors and old memories to navigate the world. Patients with cognitive impairments in new settings or environments may therefore be more confused.

The Role of Exercise

The role of exercise in both prevention and restoration of degenerative brain diseases has been highlighted by research in the last decade. Exercise, through mechanisms not yet clear, seems to help in improving brain connections, as well as “firing in the brain”. Improving core-strength has been shown to improve balance in ataxia patients. Exercise improves mood and has been shown clearly to fight depression. The sheer effect of exercise in keeping an otherwise disabled person active and outdoors makes a big change in our patient’s lives. Needless to say, all of this needs to be done in a safe environment avoiding falls and injuries.
# Johns Hopkins Ataxia Research Studies

<table>
<thead>
<tr>
<th>Condition</th>
<th>Title</th>
<th>Objective</th>
<th>Eligibility</th>
<th>Principle Investigator</th>
<th>Contact</th>
</tr>
</thead>
<tbody>
<tr>
<td>SCA 6, SCA 8</td>
<td>Ataxic Movements Study at Kennedy Krieger Institute</td>
<td>To gain a better understanding of how the cerebellum works and evaluate behavioral therapies</td>
<td>*18-85 years old</td>
<td>Amy Bastian, PhD NA_00043851</td>
<td><a href="mailto:Ataxiastudies@kennedykrieger.org">Ataxiastudies@kennedykrieger.org</a></td>
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<tr>
<td>Diagnosis of cerebellar ataxia</td>
<td>Assessment of Non-motor Functions in Cerebellar Ataxia</td>
<td>To gain a better understanding of the impact of cerebellar ataxia on cognition and emotional regulation</td>
<td>*18-75 years old</td>
<td>Cherie Marvel, PhD NA_00076574</td>
<td>Dr. Cherie Marvel, <a href="mailto:cmarvel1@jhmi.edu">cmarvel1@jhmi.edu</a></td>
</tr>
<tr>
<td>Diagnosis of cerebellar ataxia</td>
<td>Motor learning and Control in Cerebellar Ataxia</td>
<td>To gain a better understanding of the role of the cerebellum in learning and controlling skilled actions</td>
<td>*18-85 years old</td>
<td>John Krakauer, MD and Adrian Haith, PhD IRB 00036195</td>
<td><a href="mailto:krakauerlab@gmail.com">krakauerlab@gmail.com</a></td>
</tr>
<tr>
<td>SCA 1,2,3,6,7,8,10</td>
<td>Natural History Study of Genetic Modifiers in Spinocerebellar Ataxia</td>
<td>To gain a better understanding of progression rates of SCA and discover genetic modifiers that influence the age of onset and the clinical characteristics</td>
<td><em>Presence of symptoms and signs of ataxia</em></td>
<td>Chiadi Oniyike, MD and Liana Rosenthal, MD NA_00034854</td>
<td>Ann Fishman, Research Coordinator, 410-502-5816 or <a href="mailto:ann.fishman@jhu.edu">ann.fishman@jhu.edu</a></td>
</tr>
<tr>
<td>SCA 2,3</td>
<td>Magnetic Resonance Spectroscopy in Ataxia</td>
<td>To gain a better understanding and evaluate the differences between people with ataxia and healthy controls with no known ataxia</td>
<td><em>Those with early or pre-symptomatic ataxia symptoms (for 3 Tesla MRI)</em></td>
<td>Gulin Oz, MD, University of Minnesota, IRB 0502m67488; Peter Barker, D. Phil and Chiadi Oniyike, MD, Johns Hopkins University, NA_00034854</td>
<td>Diane Hutter, RN, 612-625-2340 or Ann Fishman, 410-502-5816 <a href="mailto:ann.fishman@jhu.edu">ann.fishman@jhu.edu</a></td>
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<tr>
<td>SCA 1, 3 and asymptomatic</td>
<td>Clinical Trial Readiness for SCA 1 and 3 (READISCA)</td>
<td>To gain a better understanding and assist in the design of the world’s largest group of early stage and asymptomatic SCA 1 and SCA 3 individuals or individuals with 1st degree relative who is pre-symptomatic (parent, sibling, or child)</td>
<td><em>18-65 years old</em></td>
<td>Chiadi Oniyike, MD and Liana Rosenthal, MD IRB00166215</td>
<td>Ann Fishman, Research Coordinator, 410-502-5816 or <a href="mailto:ann.fishman@jhu.edu">ann.fishman@jhu.edu</a></td>
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Ataxia Digest

Ataxia Programs and Events

In addition to the Ataxia Center’s 10th Anniversary Research Symposium and Celebration, several other social and educational programs took place in 2018. The annual Ataxia Sailing Day and the annual holiday party were well attended the weather held out as usual just for us. We also tried out kayaking (and some went water skiing) with the Baltimore Adapted Recreation and Sports (BARS) followed by a picnic which was a lot of fun at the Rocky Point State Park in Maryland. If you would like to know more about BARS programs that include adapted canoeing, sailing, kayaking, snowboarding, bowling and more please visit their website at http://www.barsinfo.org/kayaking--canoeing.html.

Educational programs included presentations and support groups to include Genetics and Ataxia; Ask the Doc, Ask the Lawyer; a care partners’ support group and presentations to the Johns Hopkins University Ataxia Ambassadors Club by various ataxia neurologists and ataxia center’s clinicians.

Upcoming Events, Support Groups and Educational Programs

We have many events and programs scheduled in 2019 and some are listed below:

Ask the Physical Therapist, Occupational Therapist and Speech Language Pathologist; Care Partner’s Support Groups; Better Understanding the Cognitive and Mental Health Issues Associated with Ataxia; Ataxia Sailing Day; Tai Chi for Individuals with Ataxia and Their Care Partners and the annual Holiday Party for Individuals with Ataxia and Their Care Partners.

For more information contact Donna at ddeleno1@jhmi.edu or 410-616-2811

New Facebook Page for Younger Persons with Ataxia

A Johns Hopkins Ataxia Center patient, with the help of the National Ataxia Foundation, created a Facebook page for people with ataxia under the age of 30 years old who can connect with each other. This group has grown tremendously under her leadership. They recently hosted a virtual meeting and support group as well. To become a member you must be between the ages of 16 to 30 years, have ataxia (not a parent or son or daughter please) and a Facebook account. The name of the group is “Under 30 with Ataxia.”
Welcome to our Ataxia Center at Johns Hopkins! The first step in the process to becoming one of our patients is to have neurology records sent to us. Please include demographic information (so we know who to contact when we get the records), neurology clinic notes within the past year, reports of your most recent MRI, lab results, and any genetic testing results. These notes can be faxed to 410-630-7900; Attn: Ataxia Center for review by one of our physicians. The decisions to accept a patient into our clinic is based on our neurologist’s assessment of whether the patient would benefit from being seen by physicians and therapists with an expertise in neurodegenerative cerebellar ataxia. Based on review of the clinic records, patients may also be scheduled with a physical therapist, occupational therapist, speech therapist, genetic counselor and for vestibular testing, neurocognitive testing, and/ or neuro-ophthalmology. Our center believes in a multidisciplinary approach to recognize and treat cerebellar ataxia. All of these of appointments are geared towards diagnosing and providing treatment recommendations. Each appointment provides a thorough work up and concentrated care to our patients.

When coming to your appointment day, please make sure to have a copy of the most recent MRI on a CD, and questions to ask your physician. Before you leave the appointment please make sure you have all referrals, orders, prescriptions or refills placed for you. Right after the visit please make sure you call to get a follow up appointment right away, since we tend to book up quickly. We always look forward to assisting in your care!

- Jana Arreola, Ataxia Clinic Coordinator

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**Medical Director**
Liana S. Rosenthal, MD

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Please consider supporting our center!

The work of the Johns Hopkins University Ataxia Center would not be possible without the generous support of the Gordon and Marilyn Macklin Foundation, the National Ataxia Foundation, our patients and the community.

For more information about supporting the center, please contact the Development Office at **443-287-7877**.