

Letter from the Director



Liana S. Rosenthal, MD, PhD

Dear Ataxia community,

When I am speaking with all of you, whether in clinic or at the many events organized by our Health Educator, I often hear about the many things that you have lost due to having ataxia. This loss is felt deeply and impacts many if not all aspects of your life. At the same time, I am amazed by your strength, resilience, and the choice that many of you make to focus primarily on what you can do, rather than what you cannot. One of the most important principles of being able to optimize your quality of life is energy conservation and compensatory strategies. This can mean many things: It may mean having to choose which holiday parties or dinners you attend, ordering more things online rather than going to the store, or allowing yourself more time to get ready to leave the house. It may also mean having friends or family wrap presents for you or having people to your house instead of going out and getting help with bathing and dressing. Overall, it means being more deliberate in your choices and plans to ensure that you have the energy to do the things are most meaningful to you.

There are a wide range of devices that can help you with energy conservation and compensatory strategies. These include button loops to help with buttoning clothes, hands-free shoes with a back that pops up or a zipper, and adaptive makeup with larger or heavier handles to help with application (for the latter, search under makeup for people with tremors). There are also a wide range of kitchen and eating devices including special knives and special silverware. Patients sometimes ask if using this assistance means they are not optimizing their independence. I would argue that these things allow for energy conservation, and therefore optimization of what really matters to you. Occupational therapists specialize in helping patients with their energy conservation and compensatory strategies and your physician and other allied health care team members are also happy to help you in this area. We look forward to partnering with you to help you be as active and engaged in the world as possible.

Sincerely,

Liana S. Rosenthal, MD, PhD

Upcoming in-person events!

* Our annual holiday party will be held on Saturday, December 9 at The Meeting House in Columbia, Maryland.

Ataxia and Adaptive Equipment

By Daniel Tekeste, OTR/L

Living with ataxia can be challenging, as everyday tasks like walking, eating, dressing, and completing home chores may become difficult. Ataxia can lead to decreased limb and body coordination, and thus may make it difficult to produce precise movements. It can also make it difficult to maintain balance when completing daily activities. Occupational therapists (OTs) can assist people with ataxia in identifying ways to adapt daily tasks to suit their individual abilities. They may also recommend and train in using various assistive devices and durable medical equipment to increase people's ability to safely and independently participate in functional activities. Below are some adaptive equipment options that people with ataxia can consider in collaboration with an occupational therapist:

Adaptive Utensils for Self-Feeding:

Weighted utensils: These eating utensils have added weight, which can help stabilize tremors and make self-feeding easier.

Plate guards: Plate guards are placed around a plate and help prevent accidental spillage of food when attempting to scoop. These devices are beneficial for people with hand tremors.

Adaptive Tools for Dressing:

Button hooks: They help make it easier to button buttons, particularly for individuals with limited fine and gross motor coordination.

Elastic shoelaces: They convert lace-up shoes into slip-on shoes.

Adaptive Devices/Software for Electronics:

Large-button phones and remote controls: Easier to use for individuals with fine motor challenges.

Dictation features and applications: If typing becomes impossible, there are software and applications that allow people to dictate texts.

Specialized Writing Tools:

Weighted pens or pencils: Reduce tremors and enhance control during writing tasks.

Writing slopes: Provide a tilted surface for writing, reducing strain on the wrist.

Specialized Cooking Supplies:

Specialized cutting boards: There are various types of specialized cutting boards. Discussing with an occupational therapist to determine the right type would be beneficial.

Electric chopper and multi-use slicer: Can be used to chop food as opposed to using a knife.

Finger guard for cutting: Protects fingers from getting cut with using knives to cut food.

It is important to note that these are just a few of the many adaptive and durable medical equipment that are available. The specific type of equipment people with ataxia will benefit from is dependent on the functional task with which the person is having difficulties, and the type and degree of physical impairment. In order to identify the right type of equipment, it would be beneficial to consult with an OT. The OT would also be able to provide resources regarding from where these items can be obtained. Most of these items can be purchased from amazon.com and/or durable medical equipment vendors/suppliers.

Mindfulness Meditation for Stress Reduction

By Neda Gould, PhD

Department of Psychiatry, Johns Hopkins School of Medicine

Mindfulness meditation is a form of present-moment awareness—just noticing what is happening inside our bodies and in the world around us as it is happening. The opposite of being mindful is being on “auto-pilot.” Mindfulness can be a very useful strategy to return to the present moment when the mind begins to get caught up in negative thought cycles and catastrophic thinking. Many research studies have shown that mindfulness meditation can reduce stress, as well as symptoms of depression, anxiety, sleep disturbance and pain.

In mindfulness meditation, we are not trying to change anything. We are simply becoming aware of what is going on without trying to judge it as good or bad. We are approaching experiences with a sense of curiosity and openness. Interestingly, when we bring our awareness to the present moment with repeated practice and allow feelings and bodily sensations to be present without trying to push them away, a natural by-product tends to be relaxation.



[Mindfulness Meditation](#)
[Brain \(c10.beauty\)](#)

Here are some tips on how to incorporate mindfulness into your day:

Take a three-breath break. Pause several times during the day and bring your attention to your breath for three breath cycles. There is no need to change your breathing in any way, simply notice that you are breathing.

Listen to a guided meditation. You can download an app on your phone or find a meditation on the internet.

Bring attention to your senses. Our senses (sight, smell, hearing, taste and touch) always reside in the present moment. Take a few minutes to become aware of what you are sensing in each of these domains. Describe what you are experiencing to yourself in a few words.

Eat mindfully. See if you can eat one meal or snack each day without doing anything else at the same time. Put down your newspaper or book. Don't have a conversation with others if possible. Simply focus on your meal. Try putting your fork down in between bites to really savor each bite. What do you notice?

Drop the story. Much of our stress comes from the “story” we create in our minds of how things should be, how they will be in the future, or how they were in the past. See if you can notice when your mind is traveling beyond the “facts” of what is true and is creating this story. See if you can bring yourself back to the facts of what is true. State the facts and drop the story.

Becoming mindful takes a lot of practice. Just remember that you only need to focus on one moment at a time. Just this moment.

How the Cerebellum Affects Vision

*By David Rastall, DO, PhD, Vestibular & Ocular Motor Oto-Neurologist,
Johns Hopkins School of Medicine*

The cerebellum is the brain's "little brain" and learns and adapts daily to make fine adjustments to limb movements, thoughts, emotions, and even sensations received from the rest of the brain. One of its critical functions is fine tuning our sense of where we are in space and then controlling our eyes to see the moving environment around us. Dysfunction of this system causes our eyes to move differently resulting in double vision, nystagmus, oscillopsia, or confusion of where we are in space, vertigo.



DIPLOPIA



These abnormal eye movements and spatial sensations are a vital part of ataxia diagnosis, as frequently (but not always) eye movements are the first definitive abnormalities in ataxia and can prove cerebellar dysfunction with a bedside exam more accurately than MRI. These subtle findings may be overlooked as identifying them requires an understanding of both dizziness and eye movement disorders.

The following eye movement problems and vertigo that are frequently overlooked with ataxias include:

Oscillopsia- is the experience of having your vision bounce or jerk around and can be caused by nystagmus, when your eyes bounce around. There are specific patterns of nystagmus that are common in cerebellar disorders.

Double vision or diplopia is also common with ataxia. It may frequently be missed as when images are double, but overlapping they can appear 'blurred' and patients with visual blur are assumed to have a refractive (i.e. glasses) problem. The most common types of diplopia with ataxia are convergence insufficiency and divergence insufficiency. To look at near objects both eyes must move towards the nose (cross) and in convergence insufficiency, they don't do this enough. The result is double images or "ghosting" at near vision. Patients with this will see double vision or visual blur when looking at near objects. To tell if the vision changes are from being near or far sighted or actual double vision, cover one eye. If it improves then this is a sign that it is alignment of the eyes, rather than focus of the eyes that is causing the problem. These visual disorders can be treated by patching one eye or with prisms, but the prisms must be expertly fitted or they will cause problems rather than solving them.

Vertigo, or the sense of moving when you are not, is also a common symptom in ataxia. This motion may be spinning, but may also be rocking back and forth, swaying side to side, feeling all types of motion like being on a boat. This type of vertigo arises from the inability of the cerebellum to integrate spatial senses into a complete spatial awareness.

With any ataxia there are some things to avoid. Meclizine (antivert) is a medication that decreases the vestibular sense from the ear to the brain. It will make dizziness and vertigo worse in patients with ataxia, as their cerebellum needs MORE information, not less to calibrate itself properly. Similarly, bifocals, progressive lenses, and "mono vision" cataract surgeries can cause issues for patients with ataxia. Bifocals work by magnifying vision when the eyes look down. When people wear these when walking they magnify the ground and end up sending false information to the cerebellum, resulting in an increase in ataxia and falls. A solution for the many adults that need both near and far vision is to wear two sets of glasses and change between them, or to wear contacts and put on readers to read, taking them off to walk.

If you think you are experiencing any of the above problems, you can reach out to see a neurologist trained in these disorders here at Johns Hopkins including David Rastall, D.O., Ph.D. and colleagues.

Eye Exercises for People Living with Ataxia

By Jennifer Millar, Physical Therapist, Johns Hopkins School of Medicine

Individuals with cerebellar ataxia may experience eye movement difficulties affecting their ability to function in daily life. Symptoms may include 1) “bouncy vision” (also called oscillopsia), 2) double vision, and 3) difficulty with eye tracking/reading.

The first step towards management of eye movement symptoms is to see an eye movement and vestibular physician, who will perform an exam and may recommend medication, special glasses, or rehabilitation. Neuro-vestibular therapy specialists work together with you and your physicians to identify the symptoms that are most impacting your daily life. To find a vestibular therapist, the Vestibular Disorders Association has a directory:

<https://vestibular.org/healthcare-directory/>

Gaze stability exercises may be prescribed in attempt to optimize the brain’s ability to predict and coordinate an appropriate motor output to maintain fixation on a visual target with head movement. Here are two exercises you can try.

Gaze stability exercise 1: Start in a sitting position and then progress to a standing position. Hand-hold a post-it with an inch sized “X” while deliberately turning your head side to side (about a 20-degree amplitude) (**Fig 1A**). Next, try moving your head up and down (**Fig 1B**), while maintaining visual fixation on the “X” target during both tasks. If the target becomes blurry, then slow the head movement down until the target is clear or try changing the distance of the target (i.e., position the target on the wall).

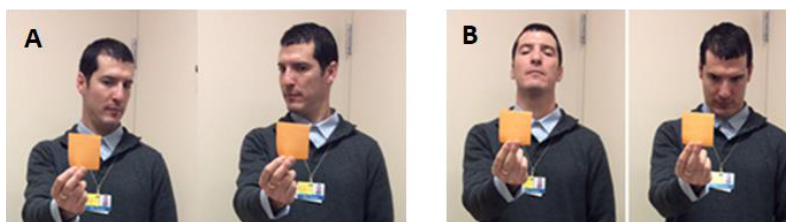


Figure 1: (A) Side to side and (B) up - down head movement when visually fixating on an X.

Gaze stability exercise 2: Hold 2 targets in front of you, with an “X” in your left hand and a “Z” in your right hand (or mounted on the wall). First, focus on the “X” with your head turned to the “X” (**Fig 2A**); Without moving your head, change your focus to the “Z” (**Fig 2B**); then turn your head towards the “Z” (**Fig 2C**). Repeat.

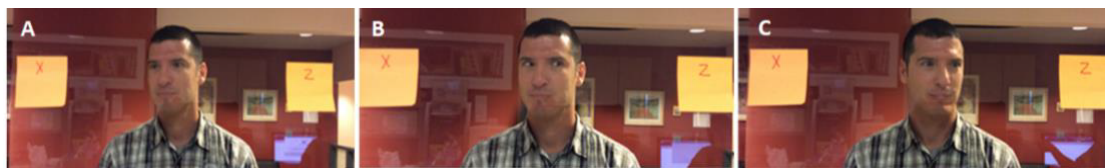


Figure 2: Visual fixation between 2 targets.

Symptoms associated with eye movement problems can negatively impact a person’s daily life. With proper medical and vestibular physical therapy treatment possibly including the exercises above, you may see gradual improvement.

Johns Hopkins Ataxia Research Studies (Current as of 10/1/2023)

Condition	Study Name	Eligibility/Information	Principle Investigator
Ataxia	Transcranial Direct Current Stimulation (tDCS) to augment dysarthria treatment in neurodegenerative ataxias IRB00239380	10 sessions of free speech therapy; 5 sessions combined with sham Age 18–80 years old Right handed Fluent speakers of English	Rajani Sebastian, PhD Contact Sarah Cust, SLP scust1@jhmi.edu 410-502-2445
Ataxia	Natural History Study of Genetic Modifiers in SCA NA_00034854	Positive genetic testing either in participant or family for SCA 1,2,3,6,7,8,10 Blood sample, neurological exam, and other tests; study visit every 12 months Ages: over 6 years old	Chiadi Oniyike, MD Liana Rosenthal, MD, PhD Contact Vanessa Nesspor vjohns23@jhmi.edu 410-616-2815
Ataxia and MSA	Biomarkers for ataxia and Multiple System Atrophy IRB00205116	Cerebellar ataxia (of unknown etiology) with symptoms for at least 8 years or MSA diagnosis Blood draw, lumbar puncture, cognitive testing 1 visit with possible yearly follow ups	Liana Rosenthal, MD, PhD Contact Vanessa Nesspor vjohns23@jhmi.edu 410-616-2815
Ataxia	Multimodal Bio-Signal Repository for Parkinson Disease and Movement Disorder IRB00234370	Established diagnosis of ataxia or other movement/neurodegenerative disorder English native speaker 1 required visit, lasting ~60-75 minutes total	Ankur Butala, MD Contact Seneca Motley Cmotley1@jh.edu 667-776-1908
Ataxia	Understanding Fatigue in Cerebellar Ataxia IRB00283000	The study will take one session (2.5 hours) You will perform a series of hand-gripping tasks and choices Age 18-80 years Fluent speakers of English	Vikram S. Chib, PhD Contact Agostina Casamento-Moran jacasame1@jhu.edu

Ataxia and vestibular	Identification of relationships of abnormal eye movements and activity in individuals with balance disorders including ataxia and vestibular dysfunction IRB00246479	This study aims to understand the relationships of kinematics during gaze, balance and walking tasks, perception of symptoms, and vestibular function in people living with ataxia. Eligibility: Diagnosis of ataxia Ambulatory, without a device Age 18–80 years old English native speaker 1 session, 2-3 hours	Jennifer Millar, PT Contact Jennifer Millar jmillar1@jhmi.edu
Ataxia	Ataxia Clinical Research Registry IRB00191999	Anyone who is seen at the Ataxia Clinic Will serve as a recruitment database and a clinical data database No additional visits are required	Liana Rosenthal, MD, PhD Contact Melissa Egerton Megerto2@jhmi.edu
Ataxia	Using Motor Imagery and Machine Learning-Based Real-Time fMRI Neurofeedback to Improve Motor Function in Cerebellar Ataxia IRB00281329 and IRB00300264 and NCT05436249	To use MRI and motor imagery to improve motor function in cerebellar ataxia Ataxia and health controls: 18–100 years old Diagnosis of SCA or cerebellar ataxia	Cherie Marvel, PhD Contact Cherie Marvel, PhD Cmarvel1@jhmi.edu
Ataxia	Cerebellar Involvement in Cognitive Sequencing IRB00328214	To use fMRI to determine how the cerebellum is involved in cognition Age 18–75 years old Fluent English speaker Confirmed spinocerebellar ataxia diagnosis	John Desmond, PhD Contact Rida Saeed rsaeed5@jhu.edu 410-502-2150
Ataxia	Monetary decision-making task to study reward perception in Cerebellar Ataxia IRB00283000	The study will take one session (2.5 hours) Age 18–80 years old Fluent speakers of English	Vikram S. Chib, PhD Contact Joonhee Lee jlee552@jhu.edu

OTHER RESEARCH RESOURCES

Clinicaltrials.gov is a registry and results database of publicly and privately supported clinical studies of human participants conducted around the world.

Connecting Organizations for Regional Disease Surveillance (CORDS) <http://www.cordsnetwork.org> Non-Governmental Organization comprised of six international networks, working to reduce and prevent the spread of infectious diseases by exchanging information between surveillance systems globally.

National Ataxia Foundation <http://www.ataxia.org/> Dedicated to improving the lives of persons affected by ataxia through support, education and research.

Fredreich's Ataxia Research Alliance (FARA) <http://www.curefa.org/index.php> National, public, 501(c)(3), non-profit, tax-exempt organization dedicated to the pursuit of scientific research leading to treatments and a cure for Fredreich's ataxia.

The Johns Hopkins Ataxia Center: How to Become a Patient in Our Clinic

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 genetic testing results. These notes can be faxed to 410-367-3318; 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 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!

- Teshome Wubishet, Ataxia Clinic Coordinator

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Please consider supporting our center!

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

For more information about supporting the center, please contact Kaylin Kopcho, Director of Principal Gifts at **443-287-7877** or **kaylin.kopcho@jhmi.edu**.

If you prefer not to receive fundraising communications from Johns Hopkins Medicine, please contact us at 1-877-600-7783 or FJHMOptOut@jhmi.edu. Please include your name and address so that we may honor and acknowledge your request.