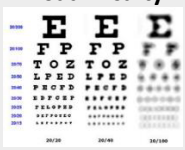
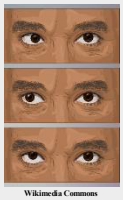
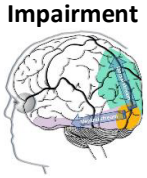
Visiting your Ophthalmologist: KAND Vision Exam List

KIF1A Associated Neurological Disorder (KAND) is a progressive neurodegenerative disorder caused by compromised axonal transport by the KIF1A motor protein. 83% of patients with KAND experience some level of visual impairment[1]. Dr. Aliaa Abdelhakim, MD, lead ophthalmologist in the KOALA clinical endpoint study, has compiled a list of potential vision symptoms in KAND, signs to look out for, and relevant ophthalmic tests. Visit our Vision in KAND page for more information.

Because KAND is a neurodegenerative disorder, it can be helpful to follow with a neuro-ophthalmologist who specializes in neurological contributions to visual deficits and can test visual processing in the brain. If KAND patients have reduced vision, a referral to a Low Vision Specialist should be considered to address visual disability using directed functional adjustments and low vision technologies.

1 Boyle et al. 2021, HGG

Clinical Feature	Signs	Tests and Considerations
Visual Acuity 	• Can't recognize people at a distance • Can't focus on nearby objects • Squints at light sources *"He can see the moon… but he cannot identify us farther than 3-4 meters."*	Patients with KAND should be checked for **Refractive Errors** such as hyperopia ("far-sightedness"), myopia ("near-sightedness"), or astigmatism. These issues can be corrected with a glasses prescription. Children tend to want to take frames off, so it is important to find comfortable frames. Resilient frames and lenses are also recommended for patients who are prone to falls, to prevent further injuries.
Field of Vision 	• Doesn't react to objects/events from side or center • Night blindness *"He is completely night blind… the combination of mobility difficulties with the blindness makes things extremely difficult for any independence safely."*	Field of vision is how much you see in your peripheral vision or "side vision". Your doctor can perform a test called **Visual Field Testing**. Visual field testing can be accomplished in different ways. Not all patients will be able to do this test as it requires focusing on the examiners face while reporting whether they can see objects in their peripheral vision.
Strabismus  Wikimedia Commons	• Eyes don't align when looking at objects (may close one eye often) • Misjudges distances • Struggles to re-orient • Tilts or turns neck to direct small visual adjustments • Pain from eye strain or headaches	Many patients with KAND have strabismus, which means the two eyes are not working together the way they should. The Oculomotor, Trochlear, and Abducens nerves control the muscles that move your eyes, and their dysfunction can cause different types of improper movement. This condition can be assessed with an **Ocular Motility Test** and be treated with glasses, patching or surgery. Follow up frequency will depend on the degree of strabismus and its impact on vision.
Optic Nerve Atrophy 	• Blind spots • Blurry vision *"Optic nerve atrophy was the first sign that something was not right."* *"He was also later diagnosed with optic nerve atrophy which has now declined to the point the ophthalmologist said it is the lowest reading the machine calculates so it is going to be difficult to track any more decline."*	The majority of patients with KAND have optic nerve atrophy[1]. Your doctor will check for optic atrophy by doing a **dilated eye exam**, but this may not detect subtle nerve atrophy. To best track exam-to-exam changes, an **optical coherence tomography (OCT) scan** would be useful. This test measures the thickness of the optic nerve. Patients who are too young, unable to focus on the target on the machine, or severely affected vision may be unable to do the OCT exam. Optic nerve function may be compromised even when the structure appears intact. Functional testing can be performed with a **Visual Evoked Potentials (VEPs)** test, which uses electrodes to check electrical communication between the eyes and the brain. This is a way to functionally assess the optic nerve function and check for neurological contributions. Not all ophthalmologists can test **VEPs**, but it may be worth asking if it is available.
Color Vision  Johannes Ahlmann, CC 2.0	• Uses the wrong colors when drawing • Lumps different colors together	The majority of assessed patients with KAND have color vision deficits. Your doctor can check for this using a **color plates booklet**, but if the patient is too young or if the vision is too severely affected, they may not be able to participate in this testing.
Cerebral Visual Impairment 	• Struggles to recall names of objects • Struggles with cluttered environments • Doesn't recognize faces • Delayed reactions to visual stimuli • Preference for moving/stationary objects *"He struggles to find an object, even when it is right in front of him."*	Visual processing goes far beyond the eye; problems with the brain's visual systems may manifest as blind spots or blurriness, or affect the way a person interacts with visual stimuli in more subtle ways. About 25% of patients with KAND have been diagnosed with cerebral visual impairment[1], which is likely underdiagnosed due to a lack of testing.