The Eye Exam & Autism Spectrum Disorders

Adaptations to the Comprehensive Eye Exam for Serving Patients with Autism Spectrum Disorders (ASDs)

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The Autism Spectrum

- Autism Spectrum Disorders (ASD) are developmental disorders that persist over the affected individual’s lifetime.

- Defining features of ASD are:
  - Social communication deficits
  - Atypical, repetitive behaviors and fixated interests

- ASDs are truly a *spectrum* and vary widely across the individuals affected.
As of May 2013, the Diagnostic and Statistical Manual of Mental Disorders (DSM-5) has defined ASD diagnostic criteria in two categories:

- **Social communication deficits**
  - Deficits in social-emotional sharing
  - Deficits in verbal and nonverbal communication, such as eye contact
  - Difficulty understanding and developing social relationships

- **Repetitive behaviors and fixated interests**
  - Stereotyped motor movements
  - Adherence to rigid routines; difficulty accepting small changes
  - Hyper- or hypo-reactivity to sensory input or unusual sensory interests
  - Intense preoccupation with unusual objects
DSM-5

- DSM-5 defines ASD as a true spectrum
- In previous DSM editions, subcategory diagnoses were individually recognized, for example
  - Asperger Syndrome
  - Pervasive Developmental Disorder-Not Otherwise Specified (PDD-NOS)
  - Rhett Syndrome
  - Childhood Disintegrative Disorder
- In DSM-5, the diagnosis of ASD encompasses all except for Rhett. Clinically, sometimes it is difficult to determine where one sub-category ends and the next begins, so the broader label seemed more appropriate to use.
A patient with ASD

- On one end of the spectrum, your patient could be
  - Nonverbal
  - Prone to self-injurious behavior, aggression, or property destruction

- At the other end of the spectrum, your patient could be
  - Fluent, but incapable of having a conversation
  - Hyperactive
  - Prone to abnormal rate, rhythm, and intonation of speech
Prevalence of ASDs

- Males are affected more often than females, at almost a 5:1 ratio\(^2\)
- 1 in 68 children were diagnosed with ASD in 2010\(^2\)
- The true cause of ASD is unclear, but genetic predisposition in combination with environmental factors likely contribute\(^2\)
- Patients with some single gene disorders can also have ASD. Examples are Down Syndrome, Fragile X Syndrome, Tuberous Sclerosis, neurofibromatosis and Smith-Lemli-Opitz syndrome.
Patient Profile

- IQ can vary from normal to in the range of intellectual disability (ID). About 60% of individuals with ASD fall into the latter category.\(^2\)

- Seizures develop in about 25% of patients with ASD\(^3\)
  - Of the ASD patients with ID, 50% develop seizures
  - Seizure activity usually begins around puberty, but sometimes begins in early childhood
  - Avoid using toys with flashing, strobe-like lights with patients with a history of seizures

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Lack of eye contact and lack of facial expression is common in ASD

Atypical language use in speed, rhythm, intonation, volume, and pronoun reversals may be noted

Repetitive physical movements without apparent purpose, known as stereotypy, are also common
  - Hand flapping
  - Jumping
  - Arm twisting
One or more of the patient’s 5 senses may be hypersensitive or dulled.

This may worsen with pain or inflammation.

Patients may cycle through hyper- then hypo-sensitivity in any of the 5 senses through time.

Patients may not be able to locate discomfort correctly.
Before the Exam Starts

- Familiarization visits to the office building and exam room can help calm the patient for the eye exam.
  - Caregivers may show the patient pictures of the office and the eye doctor from the practice’s website prior to the visit.
  - Using social stories that illustrate a visit to the eye doctor, and that use the patient as the main character, can help the patient’s familiarity with the exam.

- Some patients with ASD can be sensitive to odors and sounds.
  - The doctor and staff should avoid wearing perfume.
  - Sounds in the office should be minimized.
Before the Exam Starts

- Dr. meeting patient and care giver.
- Caregiver informed staff that patient takes time to warm up to strangers, does not like doctor offices and is very careful about personal space.
- Doctor meets patient informally and respects personal space.
Exam Flow

- The patient can be made more comfortable by minimizing wait time, both before the appointment, during the exam, and during dilation.
  - For example, scheduling the patient as the first exam in the morning or afternoon can minimize wait time before the appointment.
  - Patient medical and ocular history should be gathered and reviewed beforehand to minimize exam duration and conversation during the exam.
  - These patients may need a larger block of exam time in the schedule.
  - Allow the patient breaks when needed.
Philosophy of the Exam

- The exam should start by making the patient as comfortable as possible.
- The doctor may need to adjust his or her routine and exam flow to fit the needs of the individual patient.
- It may be helpful to modify the normal order of testing.
- In essence, the doctor is responsible for obtaining the patient’s ocular data regardless of the level of cooperation of the patient.
- Removing the “white coat” can avoid potential patient distress, especially in patients who have had many doctors visits.
- Increase safety for yourself and the patient by avoiding excessive jewelry or neck ties that may be grabbed by the patient.
Communication

- Approach the patient slowly and calmly
- Smiling and positive feedback to support your patient are important
- The patient may use verbal language, pictures, or an electronic device to communicate
- Caregivers who are most familiar with the patient can help facilitate communication between you and the patient
- Minimal conversation by the doctor and others may best preserve the patient's visual processing and attention for the best measure of acuity
Visual Acuity (VA)

- Even if the patient is verbal, using matching tasks to take visual acuity may be easier for the patient
  - Forced choice method may help obtain the VA

- Binocular acuity (both eyes open) should be the first acuity taken
  - Binocular acuity gives a functional evaluation of vision
  - The patient may not tolerate an occluder over an eye

- When taking monocular acuity, hold the occluder over the patient’s eye without crossing his/her midline. The patient will be more comfortable this way.
  - A caregiver’s hand used as the occluder may increase comfort and cooperation in some patients
  - Take advantage of any strategies that caregivers may have to elicit responses and participation from the patient
This patient is holding a comforting stuffed animal and a familiar caregiver is holding the occluder rather than the doctor.

Acuity with both eyes open would have been the initial measurement.

Patient is pointing to a matching symbol at near from a choice of at least two.
Visual Acuity Continued

- Lea symbols, Patti Pics, and numbers are great optotypes to use, according to the patient’s ability.

- For nonverbal patients, preferential looking methods are useful for obtaining an acuity:
  - Cardiff Cards
  - Teller Acuity Cards
  - Grating Paddles

- Using an isolated optotype during visual acuity can facilitate a faster and more accurate descending method to threshold acuity:
  - Whole line acuity can then be attempted at threshold.
  - Remember that significantly different acuities in isolated versus crowded optotypes are associated with amblyopia.

http://visionkits.com/products/lea-symbols
Entrance Testing

- Model the testing procedures on yourself or a caregiver to show the patient what to expect
  - Model any lights and lenses to be used
  - Even a transilluminator light may be painful to a patient with hypersensitivity. Use the lowest light settings necessary.

- Pupils
  - Constriction to light may be sluggish (longer latency, smaller amplitude, and lower velocity in pupil constriction) in patients with ASD.
  - Anisocoria may occur with fatigue. Patients with ASD may have a larger range of “normal,” but pathology must always be ruled out.
  - Remember that tricyclic antidepressants (TCAs) and antipsychotics may cause mydriasis, accommodation interference, and ocular dystonias.
Entrance Testing Continued

- **Motility**
  - Begin acuity and ocular motility testing by ensuring the patient is fixating first.
  - Fixation can often be abnormal and therefore the patient may not engage in smooth pursuit.
  - Use a caregiver’s help in suggesting the most engaging object for the patient to fixate.
  - Saccades may be abnormal in patients with ASD,\(^4\) which may affect acuity and ocular motilities.
  - Smooth pursuits may appear step-wise (saccadic) due to reduced pursuit gain.\(^8\)
Entrance Testing Continued

- Cover Test (CT)
  - Observe the patient’s alignment during all interactions with them. Strabismus may be detected simply from observation.
  - Again, use an interesting object to engage fixation. Incorporate an accommodative stimulus or object of interest to engage the patient at near.

- Other methods to assess alignment
  - Hirschberg test is used at near with eye care provider observing symmetry of light reflex in pupil.
  - Krimsky test is the Hirschberg test with the application of prism to equalize the asymmetry.
  - Bruckner Test, with simply the direct ophthalmoscope, can be a powerful detection tool for patients who do not tolerate objects near their faces. May also detect internal ocular health issue or significant refractive error.
Entrance Testing Continued

- Example of Krimsky with aid of mother.

- Color Vision (CV)
  - Can be done binocularly to save time and patient tolerance
  - Monocular color vision should be taken if pathology is suspected
  - Waggoner’s CV Testing Made Easy is useful for children who do not know numbers

http://www.testingcolorvision.com/overview.php
Entrance Testing Continued

- Near Point of Convergence (NPC)
  - Convergence Insufficiency is often associated with ASD⁹
  - Use an interesting target to engage fixation
  - Use the Hirschberg reflexes and gross observation as you bring a penlight closer to further elicit convergence insufficiency

- Stereopsis
  - Put the stereo glasses on yourself before placing them on the patient
  - The Random E or Stereo Smile cards are great to assess global stereopsis

http://www.schoolhealth.com

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Refraction

- Dry and wet retinoscopy are imperative objective data to gather during the exam.

- In some patients, the auto-refractor (handheld or table top) may be an option as your starting point, depending on their behavior and tolerance level.

- An assortment of different powered flippers can be used in place of a ret rack or loose lens when refracting.
  - These provide a larger window than the ret rack which is better tolerated by the patient.
  - They allow for a quick bracketing of the patient’s refractive error.

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Prescribing Glasses

- Patients with ASD may have difficulty tolerating objects on or around their faces such as glasses. The glasses should be adjusted to the patient’s face to give an ideal fit which will improve comfort.

- Amblyopic and strabismic patients need spectacles which will provide full correction and the best alignment.
Prescribing Glasses

- The decision to prescribe for mild refractive error depends on how the patient uses his or her vision. Use input from caregivers and teachers to evaluate the need for glasses.

- Lens treatment should impact findings, if it is to be prescribed
  - Consider sensory overload issues with new prescriptions and modify accordingly
  - For example, a patient with high myopia that has never worn glasses may be most comfortable with a partial prescription
  - Similarly, caution is advised in prescribing large amounts of astigmatism in an older teen or adult when spectacles were not considered or worn in the past.
The Dilation

- The dilation process can be both very critical and very difficult for patients with ASD
  - All patients feel discomfort with dilated pupils. In patients with ASD, the discomfort can lead to obvious agitation until resolved.
  - Shorter acting mydriatics are better tolerated in patients with ASD. When acceptable, use a short-acting dilating agent. These drops are ideal for established myopic patients.
  - Cycloplegic drops should be used when hyperopia or anisometropia is suspected

- Administering the dilating drops can be very difficult for the doctor and the patient
  - Use trained and approved professionals/staff to secure the patient if necessary
  - Lie the patient down to minimize defensive postures if needed
  - If needed, apply the drops to closed eyes and then have the patient “open” to allow the drops to enter both eyes simultaneously
Patients with self-injurious behavior, especially if directed at the head and/or eyes, may be at high risk for retinal detachment and other ocular pathologies.

- An incident of eye poking or other trauma needs to be evaluated by an eye care practitioner as soon as possible.
- Changes in daily behavior may reflect changes in vision from pathology and/or injury.
- Dilation is imperative in these cases.
The Dilated Fundus Exam

- Anterior segment evaluation can be done with the binocular indirect ophthalmoscope (BIO) in patients who may not tolerate the slit lamp.

- Again, using the lowest light setting necessary as it will increase patient comfort and cooperation.

- The patient may not follow directions such as where to look during BIO.
  - Use the patient’s eye wandering to “piece together” a comprehensive view of the entire retina in your head during BIO.
  - A direct ophthalmoscope with the patient sitting upright can be a great tool for viewing the posterior pole when other views are not possible.
The Dilated Fundus Exam

- Example of using handheld slit lamp with 78D to obtain a view of retina
Exam Frequency

- In MA, patients with ID, including ASD, should be managed under an active eye care plan of an ophthalmologist or optometrist\textsuperscript{10}
  - This includes annual comprehensive eye exams
  - These recommendations apply to individuals with blindness as well

- A comprehensive eye exam at 6 months old and at 3 years old, or sooner as directed, is recommended for all pediatric patients\textsuperscript{11}

- For “at-risk” patients, including patients with genetic disorders, eye exams should be conducted annually or as directed by an eye doctor\textsuperscript{11}

- Early, preventative care is important for healthy visual development\textsuperscript{11}
Throughout the Exam

- Patients are very sensitive to your stress level
  - Speaking in a relaxed and soothing tone communicates to your patient that you are calm and helps calm him or her
  - Singing may allow you to communicate calmness to the patient during your evaluation

- Electronic chart projectors (with internet capability) are great tools for using a patient’s favorite song or TV show on appropriate websites to direct their attention
  - This may engage the patient’s fixation at distance during retinoscopy
  - This may provide a calming entertainment source during dilation

- When you are unable to maintain a safe environment for the patient and yourself, reschedule the exam
  - Having the patient back to the office multiple times will increase familiarity and cooperation
  - The opportunity to repeat testing allows you to determine consistency in the exam findings
Throughout the Exam Continued

- Patients often understand language even if they are not able to respond verbally, make eye contact, or convey facial expressions.

- Assume the patient is aware and understanding all conversation between caregivers and yourself.

- Always speak to your patient (age appropriately) as if his or her receptive language is normal.
  - Address the patient directly to include them in the conversation.
  - Describe techniques and demonstrate them calmly before conducting them on the patient.
Exams Under Anesthesia or Sedated

- When indicated, a dilated fundus exam can be performed under general anesthesia.

- Although the need for this is rare, when a thorough examination is impossible otherwise, risk factors and patient history may warrant an exam under sedation.

Examples

- If a patient has had a recent episode of self-injurious behavior with ocular trauma, a thorough fundoscopic examination is imperative.

- If a patient’s signs and symptoms reveal a high suspicion for glaucoma and reliable IOP measurements cannot be taken, sedation may be appropriate.12
References


Further Reading on Autism Spectrum Disorders

- [www.autismspeaks.org/treatment/prt.php](http://www.autismspeaks.org/treatment/prt.php)
- [COVD Autism & Vision](http://www.autismspeaks.org)
- [Review of Optometry, Treating Patients on the Autism Spectrum](http://www.autismspeaks.org)
- Autism Spectrum Disorders in Infants and Toddlers by Katarzyna Chawarska PhD, Ami Kiln PhD, MD, Fred R. Volkmar MD and Michael D. Powers PsyD. (click here)