Screening Guide for Usher Syndrome

Florida Outreach Project for Children and Young Adults Who Are Deaf-Blind
Bureau of Exceptional Education and Student Services
Florida Department of Education
2012
Screening Guide for Usher Syndrome
Preface

This kit was designed specifically to help school personnel in conducting screenings for Usher syndrome. Usher syndrome is a genetic condition that involves hearing loss, which may range in severity from mild to profound. It also includes a type of retinitis pigmentosa (RP), which is a progressive loss of vision. This kit provides information on Usher syndrome, how to conduct a screening, the actual forms needed to conduct a screening, guidelines on next steps after the screening, resource materials, and a contact list of state and national resources.

Section 6A-6.03013(6), Florida Administrative Code, for identified students who are deaf or hard-of-hearing, requires school districts to conduct screenings for students at least once between the 6th and 12th grades. Students who are at high risk may need to be screened once in middle school and once in high school.

Florida school districts have permission to reproduce the forms and put district logos on the various questionnaires. Districts are encouraged to use the information and materials in the tool kit to share with staff, students identified with Usher syndrome and their families as appropriate. Screening is only the first part in our duty as educators and support staff in providing assistance related to the educational and emotional needs of students and their families.

If you have questions or need help, contact the Florida Department of Education, Bureau of Exceptional Education and Student Services at (850) 245-0475 or the Florida Outreach Project for Children and Young Adults Who Are Deaf-Blind: (352) 273-7530, or (800) 667-4052.
Acknowledgements

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Section 1: What Is Usher Syndrome?

Introduction

Usher syndrome is a genetic disorder that involves hearing loss, ranging from mild to profound, and the progressive loss of vision due to retinitis pigmentosa (RP), a degenerative eye condition of the retina. In addition, depending on the type of Usher syndrome, a person’s sense of balance may be affected.

Sensorineural Hearing Loss

A sensorineural hearing loss is a result of nerve damage in the inner ear, which causes interference in the nerve pathways to the brain. The inner ear consists of the cochlea, the semicircular canals, and the auditory nerve. When damage to the nerve cells in these structures occurs, the individual will have a permanent hearing loss. Many things can cause damage to the nerve cells in the inner ear. Some of these include: spinal meningitis, drugs, viral infections, heredity, and syndromes such as Usher syndrome.

http://www.sciencekids.co.nz/pictures/humanbody/eardiagram.html

Diagram 1: The Ear
Different Types of Sensorineural Hearing Loss

<table>
<thead>
<tr>
<th>Type</th>
<th>Decibel Range</th>
<th>What Can a Person Hear?</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>0 – 15 dB</td>
<td>Everything: all environmental sounds and speech sounds</td>
</tr>
<tr>
<td>Mild to moderate</td>
<td>15 – 30 dB</td>
<td>Clock ticking, whispers in quiet settings, waterfall, sounds: f, th, s, z</td>
</tr>
<tr>
<td>Moderate</td>
<td>30 – 50 dB</td>
<td>Normal conversational speech with low background noise, sounds: j, m, d, b, n, ng, e, i, a, o, r, p, h, g, ch, sh, k. Can’t hear: f, th, s, z.</td>
</tr>
<tr>
<td>Moderate to severe</td>
<td>50 – 70 dB</td>
<td>Air conditioner, loud voices, difficulty in group situations</td>
</tr>
<tr>
<td>Severe</td>
<td>70 – 90 dB</td>
<td>Piano, food blender, lawn mower, telephone, dog barking</td>
</tr>
<tr>
<td>Profound</td>
<td>90-110 dB</td>
<td>Chain saw, semi truck, helicopter, train, airplane, very loud horn</td>
</tr>
</tbody>
</table>

Retinitis Pigmentosa

Retinitis pigmentosa is an inherited disorder that results in the gradual deterioration of the light receptor cells (rods and cones) in the retina. The peripheral retina is affected first. This area of the retina contains the greatest concentration of rods (150 million), which allows a person to see in dim light, have night vision, and detect outer movement. Once the rods are affected, a person may have spotty vision (scotomas), a ring of missing information (ring scotoma), or tunnel vision (outer, lower, and upper fields are decreased).

As the rods degenerate, the cones are affected. The majority of cones are located in the macular (central) part of the retina. The fovea, a spot of the retina that contains the greatest number of cones (7 million), does not contain rods. The cones allow a person to see color, fine detail, and printed words on a page. By the time the cones are affected, there are jagged spots, the pigment separates forming clumps of pigment, bone spicules, dark spots that can be seen in the inner structures of the eye, and the optic disk is pale, yellow, and waxy. The macula may become swollen and cone cells in the fovea may die. A person may have difficulty viewing objects that are colored, small print, objects presented in the central part of the visual field, or fine detail.

What Causes Usher Syndrome?

According to The Foundation Fighting Blindness (n.d.), Usher syndrome is passed to succeeding family generations through the autosomal recessive inheritance pattern. In this type of inheritance, two copies of an Usher syndrome gene, one from each parent, are required for a person to have the syndrome. An individual who has only one copy of the gene (called a carrier) will have no symptoms of the disorder.

Parents usually do not know they may be carriers of Usher syndrome until their child is diagnosed. The possible inheritance patterns that will result in Usher syndrome are as follows:

- If one parent is a carrier and the other is not, none of their children will have Usher syndrome, but all of their children will have a 50 percent chance of carrying the gene.
• If both parents are carriers, there is a 25 percent chance that each child will have Usher syndrome.
• If only one parent has Usher syndrome and the other is not a carrier, none of their children will have Usher syndrome, but all of their children will carry the gene.
• If one parent has Usher syndrome and the other parent is a carrier, there is a 50 percent chance for each of their children to have Usher syndrome.
• If both parents have the same type of Usher syndrome, all of the children will have Usher syndrome.

Note: Usher syndrome comes in at least 10 genetic types. When two people with different types of Usher syndrome have children, none of their children are likely to have Usher syndrome.

Types of Usher Syndrome

There are three identified types of Usher syndrome. Different characteristics of vision, hearing, and balance are associated with each type. (Please refer to chart included in this section of the manual: Characteristics Associated With the Three Types of Usher Syndrome.)

Usher Syndrome Type I

A person with Usher syndrome type I is typically born with a profound hearing loss in both ears, which is sometimes characterized by a corner audiogram. (See Diagram I for an illustration of a corner audiogram.) Retinitis pigmentosa causes a visual field loss, which usually is detected in early elementary years and often progresses to total blindness as an adult. In addition, a person’s sense of balance is affected. A common indication of this absence in balance is that these children usually do not walk until they are 18–24 months old.

Diagram 1: Corner Audiogram
Usher Syndrome Type II

A person with Usher syndrome type II is typically born with a moderate to profound hearing loss in both ears, which is characterized by a sloping audiogram. (See Diagram 2 for an illustration of a sloping audiogram.) Retinitis pigmentosa is usually identified during the mid- to late-teenage years and often progresses to total blindness as an adult. The person’s balance is not affected.

![Diagram 2: Sloping Audiogram](image)

Usher Syndrome Type III

A person with Usher syndrome type III (the most rare) develops a progressive hearing loss. The hearing loss often drops drastically within a 5–10-year period and is likely to become profound. Comparative audiograms over the course of several years would indicate normal to near normal hearing thresholds at first. Subsequent audiograms show mild, moderate, severe, and finally profound hearing loss. Legal blindness may occur from 20 to 40 years of age. Individuals may gradually develop balance problems.
### Characteristics Associated With the Three Types of Usher Syndrome

<table>
<thead>
<tr>
<th></th>
<th>Type I</th>
<th>Type II</th>
<th>Type III</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Hearing Loss</strong></td>
<td>Congenital severe to profound bilateral sensorineural hearing loss; corner audiogram</td>
<td>Congenital moderate to profound bilateral sensorineural hearing loss; sloping audiogram</td>
<td>Normal hearing to moderate bilateral sensorineural hearing loss at birth; rapid progression of hearing loss for 5–10 years</td>
</tr>
<tr>
<td><strong>Vision Loss</strong></td>
<td>Early onset of blind spots; early onset of night blindness; peripheral vision loss; often results in total blindness during adult years</td>
<td>Blind spots or ring scotoma by teenage years; peripheral vision loss; often does not progress to total vision loss</td>
<td>Blind spots or ring scotoma by teenage years; peripheral vision loss; progression varies but usually legally blind by young adult years</td>
</tr>
<tr>
<td><strong>Other Visually Related Conditions</strong></td>
<td>Cataracts</td>
<td>Cataracts</td>
<td>Myopia and astigmatism</td>
</tr>
<tr>
<td><strong>Balance</strong></td>
<td>Balance problems; walks at 18–24 months; does not get dizzy</td>
<td>Balance normal</td>
<td>Progressive balance problems</td>
</tr>
<tr>
<td><strong>Genetics</strong></td>
<td>1A long arm of 14; 1B-Myosin VIIA; 1C-Harmonin; 1D-Cadherin; 1-E Protocadherin; 1F-?</td>
<td>2A-Usherin; 2B-Chromosome 3p; 2C-Chromosome 5q</td>
<td>3 long arm of 3 (Finnish descent)-Novel gene</td>
</tr>
</tbody>
</table>
Section 2: Why Is Screening Important?

Screening for Usher syndrome is important for several reasons:

1. Early identification impacts medical decisions, educational decisions/services, and social implications for the student as well as the family members. 
   a. The student can receive counseling and support to prepare him or her for the future.
   b. A diagnosis of Usher syndrome allows parents and children to consider genetic counseling and testing for other children in the family.

2. Among children who are born profoundly deaf, 3 to 6 percent can be expected to have Usher syndrome type I. Among children born hard-of-hearing, Usher syndrome type II probably occurs at the same rate. Vision screenings for retinitis pigmentosa are not routinely performed in the school setting, resulting in an under-identification of students with Usher syndrome.

3. Students with Usher syndrome typically exhibit behaviors that may be misinterpreted as clumsiness, slowness, or uncooperativeness. Identification of Usher syndrome in a student prevents this type of misinterpretation.

4. Early identification can provide students and their families with access to new research being developed for the treatment of Usher syndrome and opportunities to participate in clinical studies on a national level.
Section 3: Where Do I Begin?

What Is Included in Usher Syndrome Screening?

There are three parts to a screening: audiogram review, paper screening, and on-site screening.

In the audiogram review, students who have a conductive hearing loss as documented on their audiogram do not need to proceed with further screening. If a student has a sensorineural loss, the student is a candidate for further screening.

The paper screening consists of a set of questionnaires. The paper screening identifies family history that may indicate the potential for Usher syndrome. It may also identify behavior characteristics that may indicate night blindness, a visual field loss, or blind spots.

The on-site screening consists of three parts: visual field, cone adaptation, and balance. The visual field and cone adaptation screenings are used to identify students who demonstrate visual behaviors that are characteristic of retinitis pigmentosa, rod deterioration, or cone deterioration. The balance screening is used to identify students who demonstrate balance difficulties.

Screening is only the first step in identifying Usher syndrome. For a definitive diagnosis of RP, an ophthalmologic evaluation including an electroretinogram (ERG) is required. Only an ophthalmologist can administer this test. (For more information on ERGs, see Appendix A: What Is an Electroretinogram (ERG) Evaluation?) Additionally, genetic counseling is recommended to confirm the student has Usher syndrome.

Who Conducts the Screening?

The screening team may be composed of a teacher of the deaf/hard-of-hearing, a teacher of the visually impaired, a school nurse, an audiologist, and/or other school personnel, as determined by the school district. It is important that screeners are appropriately trained and at least one member of the team is familiar with the student. When screening students who rely on sign language for communication, make sure interpreters are included in the screening process.

Next Steps

Before you begin a screening, familiarize yourself with the different types of Usher syndrome, the inheritance patterns, and why screenings are important. Next, do the following:

1. Read this entire manual and review kit materials.
2. Conduct a review of audiological reports. This will identify students who require the paper screening.
3. Copy the forms needed for the paper screening. These are found in the Forms Packet and in Appendix E.
4. Conduct the paper screening. After the paper screening has been conducted, determine if an on-site screening is required.

5. Obtain a Cone Adaptation Test.

6. Copy the forms necessary for the on-site screening.

7. If you have questions regarding the on-site screening, contact the Florida Outreach Project for Children and Young Adults Who are Deaf-Blind at (800) 667-4052 or (352) 273-7530, or the Bureau of Exceptional Education and Student Services of the Florida Department of Education at (850) 245-0475.

8. Evaluate the results from the paper screening and the on-site screening. Determine the student’s referral status by following the instructions in Section 6. If you are not sure about the results, read the case study in Appendix C.
Section 4: How Is the Audiogram Report Screening Conducted?

Not all students who are deaf or hard-of-hearing are at risk for Usher syndrome. The review of audiological reports serves to identify students most likely to require a further screening. There are key characteristics of a student’s audiological profile that indicate the need for further screening.

A student is considered to be at low risk and does not need further screening if:

- The audiological report states the student has a conductive loss.
- The audiogram shows a unilateral, sensorineural loss.
- The slope of the loss depicted on the student’s audiogram rises in the higher frequencies.

A student is considered to be at high risk and does need further screening if:

- The student has a corner audiogram (see Section 1, Diagram 1).
- The student’s audiogram shows a downward slope (see Section 1, Diagram 2).
- The student’s past audiograms show a steady pattern of hearing loss over a period of 5–10 years.

How Is the Paper Screening Conducted?

**Questionnaires**

Questionnaires are used to gather more information about the student’s family and health history, visual functioning, balance, and adaptation skills in school and social situations. The family, school personnel who are familiar with the student, and the student (via interview) are to complete questionnaires. Questionnaires in Appendix E and the Forms Packet include the following:

- Family History Questionnaire
- Student Questionnaire
- Visual and Motor Behavior Questionnaire

**Notification**

A letter of notification must accompany the Family History Questionnaire. Although this is not a permission slip, it should include information regarding the parent’s right to refuse screening for their child. Refusal for any health screening should be submitted in writing in accordance with subsection (7)(e) of s. 381.0056, F.S.

If the student is at low risk for Usher syndrome, the family should receive written notification. A sample parent notification letter is in Appendix D.

The individual educational plan (IEP) team should receive notification of the upcoming screening and be informed of the results.
Family History Questionnaire

When reviewing the Family History Questionnaire, please note the following items. The team should continue with the screening process if:

- The student has not been diagnosed with another syndrome.
- The student did not have multiple organs affected at the time of birth or as a result of a degenerative condition.
- The student’s family history indicates no multiple generations of hearing loss, or neither biological parent is deaf or hard-of-hearing.
- The student does not show signs of intellectual disability.
- The student did not walk until 18 months or after.
- The student was born deaf or hard-of-hearing (bilateral sensorineural loss). Or the student was born with normal hearing and it declined significantly within a 5–10-year period and that decline was not the result of infection, disease, or trauma.
- The student demonstrates visual behaviors that indicate difficulty seeing at night, in dim light, or a person signing from the side of his or her body.
- The student has balance difficulties when walking on uneven surfaces, bumps into objects like tables and chairs, or knocks things over when reaching for them.
- The student has no noticeable physical characteristics (e.g., white patch of hair) or other health issues that could possibly indicate another syndrome (e.g., Refsum syndrome) that could result in deaf-blindness.

Note: If both parents are hearing or one is hearing and one is deaf AND there are one or more relatives from past generations who were born deaf or hard-of-hearing and lost their sight in their mid-teens to early 30s, the student should receive an on-site screening. If both biological parents have Usher syndrome, the student needs to receive an on-site screening.

The Visual and Motor Behavior Questionnaire

Educational service providers who are familiar with the student should complete the Visual and Motor Behavior Questionnaire. These may include, but are not limited to, teachers, therapists, paraprofessionals, interpreters, and school nurses, as appropriate.

The following are items of concern that are likely to be revealed by the Visual and Motor Behavior Questionnaire. A large number of positive responses on this questionnaire may indicate visual field losses or balance difficulty.

- The student bumps into objects, misses curb cuts, or misses the top of stairways.
- The student turns his or her head while reading or uses a finger to mark a spot on the page.
- The student does not respond to signing or waving to the side of his or her body.
- The student is not able to see cars approaching from the side of his or her body at an intersection.
- The student is not able to function in dim light or has night blindness.
- The student has problems adjusting to environments ranging from brightly lit to dimly lit.
- The student does not fully participate in groups, does not like new situations, and is the last to enter a room.
• The student is unable to ride a bike, or it took a very long time for him or her to learn.
• The student is unable to function in low-contrast situations.

**Student Questionnaire**

The following list may also indicate visual field losses or balance problems.

• The student reports tripping over objects on the floor.
• The student reports being unable to adjust to light within a couple of seconds when entering a darkened area (e.g., movie theater).
• The student reports being unable to see the stars at night.
• The student reports being unable to see moving objects from the side of his or her body (e.g., friends waving, cars moving, or individuals signing).
• The student reports difficulty riding a bicycle.
• The student reports bumping into things or knocking over objects placed on a table.

If the information from the paper screening indicates a large number of matching responses (i.e., half or more) on the Student Questionnaire and the Visual and Motor Behavior Questionnaire, or there are obvious concerns about the student, the student should receive an on-site screening.

**After You Have Reviewed all Materials**

**Timeline Chart**

<table>
<thead>
<tr>
<th>What?</th>
<th>Who Completes?</th>
<th>Time Allocated</th>
</tr>
</thead>
<tbody>
<tr>
<td>Review audiological reports and audiograms</td>
<td>Audiologist or teacher of the deaf/hard of hearing</td>
<td>Week 1</td>
</tr>
<tr>
<td>Distribute parent letter with Family History Questionnaire</td>
<td>Parents, guardians, or care providers</td>
<td>Weeks 2 &amp; 3</td>
</tr>
<tr>
<td>Complete Visual and Motor Behavior Questionnaire</td>
<td>Teachers and/or service providers</td>
<td>Week 4</td>
</tr>
<tr>
<td>Complete Student Questionnaire</td>
<td>Student with assistance</td>
<td>Week 4</td>
</tr>
</tbody>
</table>

If materials are not returned within this time, contact families by telephone to encourage their involvement in the screening process. If the parent has not refused the screening, proceed with the Student Interview and Visual and Motor Behavior Questionnaire regardless of whether the Family History Questionnaire has been returned.

Review all questionnaires to determine if the student should receive an on-site screening. In general, if the audiogram is typical of those with Usher syndrome and there are consistent concerns evidenced by the answers on the questionnaires, it is recommended to conduct further screening.
Section 5: On-Site Screening Process

Preparation

1. Set date(s) for screening in collaboration with school schedule so there are no conflicts with field trips, enrichment experiences, assemblies, or other school activities.

2. Plan for the screening day. This includes the following:

   a. Gather the following materials
      • Cone Adaptation Test
      • Appropriate rooms
      • Timers or stopwatches
      • Students’ screening folders (includes completed Family History Questionnaire, Student Questionnaire, Visual and Motor Behavior Questionnaire, student audiograms, and signed release of information)
      • Blank forms for on-site screening (cone, balance, and visual field)

   b. Arrange for interpreters to match communication needs of the students (e.g., American Sign Language [ASL], signed English, oral).

   c. Identify room(s) for each screening area (i.e., cone, visual field, and balance) and determine if free from visual and auditory distraction:
      • Cone: Can the room be made dark enough for testing?
      • Visual field: It should be a small room with good lighting and three chairs (screener, interpreter, and student).
      • Balance: It should be a room large enough for two screeners, two interpreters, and two students. Each student should be able to extend both arms out to the side of his or her body.

   d. Recruit people to assist with the screening (contact the Florida Outreach Project for Children and Youth with Deaf-Blindness; local teachers of the deaf/hard-of-hearing; local teachers of the visually impaired).

   e. Develop a schedule for screening.

   f. Inform teachers of time, day, and location of the screening to release students from class.

   g. Hold a planning meeting with all screeners to run through the screening process.
      • Identify who will work at each screening area.
      • Identify someone to assign students to screening area and give them folders when they first enter the screening area.
      • Explain how to complete the forms.
      • Have screeners practice the process on each other.

The Role of the Screener

Screeners who are identified to assist in the screening process must have adequate information about Usher syndrome, how it occurs, behavioral characteristics, and psychosocial and educational ramifications. Examples of possible district staff include speech therapists, audiologists, teachers of the visually impaired and/or teachers of the deaf/hard-of-hearing, school and/or community nurses, and medical doctors in the neighboring community.
The total number of screeners will depend upon the number of students needing the on-site screening. If only two to five students require the on-site screening, only two or three screeners will be needed. However, if more than five students will receive the on-site screening, at least one screener should be identified for each screening area.

To ensure consistency and accuracy of test results, it is essential that screeners meet in advance of the on-site screening to learn about the purpose of screening, learn their roles and responsibilities, and develop clear expectations for the screening process by practicing the various tests on each other.

**The Role of the Interpreter**

For the screening process to be valid, interpreters must be provided for students who rely on a sign language or oral interpreter in the classroom. In addition, interpreters must match the communication needs of the student (e.g., students using ASL paired with ASL interpreters; students using a signed English system paired with interpreters trained in the student’s sign language system). Likewise, if a student relies on amplification, an assistive listening device, glasses, or other visual aids, these must be used during the screening process. A child’s cognitive abilities may dictate alternative strategies to achieve effective communication.

The number of students being screened and the room layout will determine the number of screeners and interpreters. Interpreters should arrive 15 minutes before the screening process begins. The screener for each station will explain the testing process to the assigned interpreter. Breaks should be provided every 30 to 60 minutes so interpreters can refresh themselves.

**Visual Field Test**

**People and Materials Needed**

- One screener
- One interpreter
- Three chairs
- Visual Field Test Form (within student folder)
- Allow 10 minutes per student

Note: Seek assistance from the local teacher of the visually impaired and have him/her conduct a functional vision assessment, including visual field test. If there is not a teacher of the visually impaired in your area, arrangements may be made with a neighboring district or you may contact the Florida Department of Education at (850) 245-0475.

**Room Setup**

The screener and student should be seated facing each other. There should be approximately one arm’s length between them. The interpreter should stand behind the screener.

**Screener Procedures**

1. Review procedures on the Visual Field Form for more detailed instructions.
2. Explain the procedures for the Visual Field Test.
3. Explain the activity to the student.
4. Conduct the activity.
5. Document results.
6. Return the Visual Field Test Form to the student folder.

Balance Test

**People and Materials Needed**

- Screener(s)
- Interpreter(s) (depending on the number of students)
- Balance Form (within student folder)
- Allow 10 minutes per student

**Room Setup**

Unlike the Cone Adaptation Test, two students can be screened for balance in the same room. Each student should stand facing the interpreter. The screener should stand behind each student.

**Screener Procedures**

1. Review procedures on the Balance Form for detailed instructions.
2. Explain the procedures for the Balance Test.
3. Tell the student to see if he can keep his balance during the first component of the test.
4. Conduct the first component of the Balance Test.
5. Explain that the second component of the test could be more difficult than the first. Ensure the student that you will provide support if he or she loses balance or sways.
6. Conduct the second component of the Balance Test.
7. Reinforce student for completing the test.
9. Return Balance Form to student folder.

Cone Adaptation Test

**People and Materials Needed**

- Screener (may use two screeners if two separate rooms with the same light level are available)
- Interpreters (depending on the number of students)
- Allow 15 minutes per student
- Cone Adaptation Form (within student folder)

**For Each Screening Room**

- Cone Adaptation Test
- Table with dark top
- Three chairs
- Two timers or stopwatches
- Cone Adaptation Form (within student folder)
**Room Setup**

Select a screening room with lights that can be turned off but also possessing an additional, dim source of light (e.g., a night light, a light from a door or window, an outside window with blinds closed and possibly covered with sheet or blanket). The dim lighting in the room should imitate dusk or twilight. The source of light must be indirect and not fall upon the screening materials. The student and screener should be seated across from one another at a table. The interpreter should be standing next to the screener, in full view of the student.

**Screener Procedures**

1. Review procedures on Cone Adaptation Form for more detailed instructions.
2. Explain the Cone Adaptation Test.
3. Explain that when the light is turned off, the student begins the activity.
4. Conduct the Cone Adaptation Test and document results on the student’s Cone Adaptation Form.
5. Reinforce the student for completing the test.
6. Conduct the Cone Adaptation Test and document results again.
7. Reinforce the student for completing the second test. The screener may choose to share the results with the student.
8. Return Cone Adaptation Form to the student folder.

Note: Document on the screening form any irregularities that occur (e.g., chips fall on the floor; the student uses his or her hands instead of both eyes to locate chips).
Section 6: Evaluating the Results

Once you have completed the screening process, the information should be compiled for each student onto the Usher Screening Data Summary sheet. Once the form has been completed, there are three possible actions you can take.

1. Pass the student (i.e., eliminate from further screenings)
2. Re-check the student within one year
3. Refer the student for a medical diagnostic evaluation

The criteria for each of these actions are summarized below.

Pass the student if:

- The student passed all three screenings (cone adaptation, balance, and visual field).

Re-check the student in one year if:

- The student did not pass, possibly due to difficulties understanding, then consider referring on for medical diagnostic evaluation.
- The student's audiogram reflects a corner or is sloping.
- The family history, student interview, and behavior checklists are inconsistent with each other in regard to night blindness, a pattern of visual field loss, or other visual losses.

Refer for a medical diagnostic evaluation if:

- The family history indicates both biological parents have Usher syndrome, one parent has Usher syndrome, or there is a past history of hearing losses with a visual impairment.
- The student walked after 18 months of age or later.
- There is medical history that suggests acuity loss, blind spots, eye muscle imbalance, glare and contrast sensitivity, or color blindness.
- The student's audiogram reflects a corner or is sloping.
- The student did not pass the Visual Field Test or was not able to test.
- The student passed less than 75 percent of the Cone Adaptation Test within a two-minute time period, and less than 75 percent of the Balance Test.
Appendix A: What Is an Electroretinogram (ERG) Evaluation?

An ERG is an evaluation that measures the electricity discharged by nerve impulses in the retina. A board certified ophthalmologist conducts this test. It can detect retinitis pigmentosa in children beginning at six to seven years old, and for some children it can be detected at younger ages (reportedly as young as one year of age).

This test is conducted using two methods: the skin electrode method and the lens method. When using the skin electrode method, a person has electrodes attached to the skin in certain locations around the eye. These electrodes are connected to a computer that records responses of the retina. The person places his or her head in a globe called a Gonzalez Globe. Flashes of colored lights are presented in various locations in the globe for a brief amount of time.

When using the lens method, a person wears contact lenses with electrodes attached. These electrodes are connected to a computer. The person is presented with flashes of colored light in the globe, like the ones used in the skin electrode method. As in the skin electrode method, the retina’s responses are recorded. The doctor will interpret the responses that indicate retinitis pigmentosa.
Appendix B: How to Explain Usher Syndrome to Students

The following is a suggested script you can follow or adapt to explain Usher syndrome to students.

**Step 1: Briefly describe what happens with Usher syndrome**

*This will depend on the age and comprehension level of the student.*

Usher syndrome is when the part of your eye called the retina gets sick. When it gets sick, you cannot see things as well as when you were younger. Your sight may get even worse as you get older.

If you think your eyes are not seeing well, you need to tell your parents, your teacher, or an adult friend. While you are in school, you may be asked to do some activities with a teacher or nurse that will test how well you see. This is called an Usher Screening Test. There are three parts to this test.

**Step 2: Explain the Cone Adaptation Test**

The first is the Cone Adaptation Test. In the Cone Adaptation Test, you will have about 15 squares. Some are white, some are blue, and some are red. An adult will ask you to put all the red squares in one pile, all the white squares in another pile, and all the blue squares in a third pile. You will do this two times. The room will be almost dark.

When the lights are off, the adult can learn how your eyes use a little light to help you see. Some students have problems when there is only a little bit of light in the room. If you think you have trouble seeing with just a little light, do not worry. That does not mean you have Usher syndrome.

**Step 3: Explain the Balance Test**

The second test is the Balance Test. This test is done with the lights ON in the room. Sometimes your eyes will be open; sometimes they will be closed.

First, you will stand with ONE FOOT IN FRONT OF THE OTHER. Your hands will be at your side and your eyes will be open. You will lift your arms up to the side like you want to fly. The adult will stand behind you and lightly touch the sides of your body (trunk) to see if you can remain standing. Then you will do the same thing with your eyes closed.

Next you will stand WITH YOUR FEET TOGETHER. Your hands will be at your side and your eyes will be open. You will lift your arms up to the side like you want to fly. The adult will stand behind you and lightly touch the sides of your body to see if you can remain standing. Then you will do the same thing with your eyes closed. That’s it!
Step 4: Explain the Visual Field Test

During the Visual Field Test, you will sit and/or stand directly across from an adult. You will be asked to sit/stand straight and to keep your eyes on the adult’s nose at all times. The adult’s hands will be at the side of his or her body. When you see the adult's fingers wiggle, raise your hand. You will do this two or three times, then you’ll be finished! The important thing is to keep your eyes on the adult’s nose. Don’t worry if you have difficulty seeing the wiggling fingers. This does not mean you have Usher syndrome.

Remember, it is important to ALWAYS ASK QUESTIONS. If you do not understand what to do, are scared about the test, or are worried about how you did, tell the adult. Someone will be glad to help you.
Appendix C: A Case Study/Usher Syndrome I

Background: Sam Evans
Sam Evans is 14 years old. He was born with a profound bilateral hearing loss. Sam uses American Sign Language and print for receptive and expressive communication. His teachers and parents note he possibly has some difficulties with vision. Sam was referred to his school nurse for an Usher screening.

In preparation for the on-site screening, the nurse sent the Family History Questionnaire to Sam’s parents and to all of Sam’s teachers, obtained his audiogram from the audiologist, and had Sam’s homeroom teacher conduct a student interview with Sam. The results are below.

Paper Screening: Family History Questionnaire
Sam’s mother completed the form. She indicated that Sam was deaf at birth. He tried hearing aids when he was five, but they were discontinued when they did not appear to help him. She indicated that he has worn glasses since the age of seven. Additionally, she noted several behaviors that indicated Sam was having difficulty seeing at night (e.g., afraid of shadows and difficulty seeing signs or gestures in dim light) and difficulty with light adjustment (e.g., wears sunglasses and stops when exiting a darkened room to the outdoors).

She further reported that Sam:

- Did not walk until he was 20 months old
- Kept his head on the ground when he crawled
- Trips over things in front of him
- Doesn’t see the curb or the first stair of a stairway
- Holds onto someone’s hand when the lights are dim or it is dark
- Had difficulty riding a bike
- Was hospitalized for a tonsillectomy, a broken arm, and a broken hip
- Takes vitamins and Tylenol (as needed) for headaches
- Is very sensitive about tipping or bumping into things, and does not like to be in a darkened room without the lights on

Although Sam’s mother reported no problems during pregnancy, she did indicate a family history of myopia, bifocals, and an aunt who was diagnosed as hard-of-hearing during middle age. Neither Sam nor his family members have seen a geneticist.

Paper Screening: Student Questionnaire
Sam reported that his eyes take longer than a minute to adjust to a darkened room. He stated that he sometimes trips over things, spills liquids when pouring from a container into a cup, and has difficulty finding small objects dropped on the floor. He indicated that sunlight bothers his eyes. He wears sunglasses when he is outdoors.

Paper Screening: Visual and Motor Behavior Questionnaire
Both Sam’s mother and his social science teacher completed the Visual and Motor Behavior and Family History Questionnaires. There were consistent patterns in night blindness, visual fields, balance, and other areas. There were inconsistencies reported for glare, contrast, and acuity.
Paper Screening: Audiogram
Sam’s audiogram revealed a corner audiogram consistent with audiograms that would indicate Usher syndrome type I. He has a 90 dB loss that drops off dramatically in the higher frequencies.

Based on the patterns of visual behaviors, his audiogram, and family history, it was determined that Sam needed to receive an on-site screening.

On-Site Screening: Cone Adaptation Test
Sam completed both trials of the Cone Adaptation Test within 30–45 seconds. He received 100 percent correct for grouping the white squares, and less than 75 percent for grouping the red and blue squares.

On-Site Screening: Balance Test
Sam failed 75 percent of the Balance Test. He only passed the section with his eyes open and both feet together.

On-Site Screening: Visual Field Test
Sam’s visual fields were tested using both eyes. Both horizontal and vertical fields indicated a possible loss in the periphery.

Analysis of On-Site Screening:
Sam did not pass any of the on-site screening tests. His paper screening indicated visual field losses, balance difficulties, and night blindness; and his family history indicated the presence of Usher syndrome. The school nurse referred Sam for an extensive medical evaluation, including an ophthalmological evaluation with a physician experienced in visual field losses. The test was paid for by Sam’s parents’ insurance. Sam’s mother currently is waiting for the test results.
March 21, 2012

Dear Parent/Guardian:

To comply with the requirements of the Florida Department of Education State Board Rule 6A-6.03013(6), Florida Administrative Code (F.A.C.), your child will be screened this year for Usher syndrome. This screening is simple and quick to administer and consists of a preliminary Family History Questionnaire to identify those students who are at risk, followed by screenings for dark adaptation, balance, and visual field discrepancies. You may opt out of this screening by sending a letter to your child’s teacher prior to the scheduled screenings, in accordance with subsection (7)(e) of s. 381.0056, F.S.

Usher syndrome is a genetic disorder involving the loss of both hearing and vision. The screening hearing loss is congenital (i.e., occurs at birth or shortly thereafter) with a loss of vision due to retinitis pigmentosa (RP) beginning later in life. It is estimated that 3–6 percent of people with a congenital hearing loss will have Usher syndrome. By screening students who are deaf and hard-of-hearing for a vision loss, Usher syndrome can be diagnosed at the earliest possible time.

Attached is the Family History Questionnaire. Please complete this questionnaire as directed and return it to your child’s teacher as soon as possible. If the information you provide about your child’s hearing and vision does not indicate your child is at risk, no further screening will be administered. Your child will also complete a questionnaire. If your child does not pass the screenings, you will be notified.

Please note that this Usher Syndrome Screening can only be considered as a preliminary and basic screening and does not replace regular eye exams administered by an eye care specialist.

Thank you for your attention to this important issue. If you have any further questions, please contact me at _________________, or you may contact your child’s teacher or the school nurse.

Sincerely,

Attachment
March 21, 2012

Dear Parent/Guardian:

Your child was administered a preliminary screening for Usher syndrome as required by the Florida Department of Education State Board Rule 6A-6.03013(6), F.A.C. The results are as follows:

Family Questionnaire  _____ High Risk  _____ Low Risk
Dark Adaptation Screening  _____ High Risk  _____ Low Risk
Visual Field Screening  _____ Fail  _____ Pass
Balance Screening  _____ Fail  _____ Pass

This is a screening, not a test, but the results indicate that your child has high risk factors for this syndrome. It is recommended that you take your child to an ophthalmologist who will provide a more accurate and thorough examination.

Thank you for your attention to this important issue. If you have further questions, please contact me at ____________________.

Sincerely,

Teacher for the Deaf/Hard of Hearing
# Appendix E: Forms for Paper Screening

*The following forms were adapted with permission from the Kansas Usher Screening Project.*

## Family History Questionnaire

<table>
<thead>
<tr>
<th>Student's name:</th>
<th>Date of birth:</th>
<th>Today's date:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Address:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Phone number:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Name of person filling out form:</td>
<td>Relationship to student:</td>
<td></td>
</tr>
</tbody>
</table>

## Questions about the Child:

### Hearing

<table>
<thead>
<tr>
<th>How old was your child when he/she became deaf?</th>
<th>□ Yes □ No</th>
</tr>
</thead>
<tbody>
<tr>
<td>How did he/she lose hearing?</td>
<td></td>
</tr>
<tr>
<td>Has he/she ever worn hearing aids?</td>
<td>□ Yes □ No</td>
</tr>
<tr>
<td>If yes, did hearing aids help?</td>
<td>□ Yes □ No</td>
</tr>
<tr>
<td>Was your child's hearing ever better than it is right now? Explain:</td>
<td>□ Yes □ No</td>
</tr>
</tbody>
</table>

### Vision:

<table>
<thead>
<tr>
<th>Does your child wear glasses?</th>
<th>□ Yes □ No</th>
</tr>
</thead>
<tbody>
<tr>
<td>If yes, how old was your child when he/she received glasses?</td>
<td></td>
</tr>
<tr>
<td>Does your child have difficulties seeing objects far away (nearsighted)?</td>
<td>□ Yes □ No</td>
</tr>
<tr>
<td>Does your child have difficulties seeing objects close up (farsighted)?</td>
<td>□ Yes □ No</td>
</tr>
<tr>
<td>Has your child ever had eye drops put into his/her eyes to dilate them? If yes, were there any problems with the drops? Describe:</td>
<td>□ Yes □ No</td>
</tr>
<tr>
<td>Question</td>
<td>Yes</td>
</tr>
<tr>
<td>------------------------------------------------------------------------</td>
<td>-----</td>
</tr>
<tr>
<td>Does your child have difficulties seeing at night or in the dark?</td>
<td></td>
</tr>
<tr>
<td>Is he/she afraid of the dark or of shadows?</td>
<td></td>
</tr>
<tr>
<td>Does he/she have difficulties seeing the stars at night?</td>
<td></td>
</tr>
<tr>
<td>Does he/she complain that light bothers his/her eyes?</td>
<td></td>
</tr>
<tr>
<td>Does he/she need to wear glasses in bright sunlight?</td>
<td></td>
</tr>
<tr>
<td>When going from bright light to dim light, or vice versa, does he/she ever stop suddenly and look around?</td>
<td></td>
</tr>
<tr>
<td>Does he/she ever confuse colors?</td>
<td></td>
</tr>
<tr>
<td>If yes, is the problem with dark colors like navy, black, or brown?</td>
<td></td>
</tr>
<tr>
<td></td>
<td>yellow and blue</td>
</tr>
<tr>
<td></td>
<td>red and green</td>
</tr>
<tr>
<td><strong>Balance:</strong></td>
<td></td>
</tr>
<tr>
<td>At what age did he/she sit?</td>
<td></td>
</tr>
<tr>
<td>Crawl? _____ months</td>
<td></td>
</tr>
<tr>
<td>Walk alone? _____ months</td>
<td></td>
</tr>
<tr>
<td>Did he/she have any difficulties crawling or an abnormal crawl?</td>
<td></td>
</tr>
<tr>
<td>If yes, please describe how your child moved or crawled about:</td>
<td></td>
</tr>
<tr>
<td>Did he/she walk with feet apart longer than most children?</td>
<td></td>
</tr>
<tr>
<td>If yes, until what age? _____ months</td>
<td></td>
</tr>
<tr>
<td>When learning to walk, most children sit down hard on their seats and catch themselves with their hand if they start to fall. Did you child do anything differently?</td>
<td></td>
</tr>
<tr>
<td>If yes, describe what happened:</td>
<td></td>
</tr>
<tr>
<td>Did your child wear out lots of shoes or trip because he/she “shuffled” feet instead of picking up feet?</td>
<td></td>
</tr>
<tr>
<td>Question</td>
<td>Yes</td>
</tr>
<tr>
<td>-------------------------------------------------------------------------</td>
<td>-----</td>
</tr>
<tr>
<td>Do you think your child is clumsier than other children the same age?</td>
<td></td>
</tr>
<tr>
<td>If yes please describe:</td>
<td></td>
</tr>
<tr>
<td>When your child was 3–5 years old and you were going from the car to the house at night did he/she:</td>
<td></td>
</tr>
<tr>
<td>Stand or wait until someone came to take his/her hand?</td>
<td></td>
</tr>
<tr>
<td>OR</td>
<td></td>
</tr>
<tr>
<td>Walk to the house by him/herself?</td>
<td></td>
</tr>
<tr>
<td>Does he/she have problems now when walking on bumpy ground?</td>
<td></td>
</tr>
<tr>
<td>Does he/she often bump into objects like tables and chairs?</td>
<td></td>
</tr>
<tr>
<td>Does he/she knock things over at mealtime?</td>
<td></td>
</tr>
<tr>
<td>When he/she walks on a curb or four-inch-wide board, does he/she fall off?</td>
<td></td>
</tr>
<tr>
<td>Does he/she know how to ride a two-wheeled bicycle?</td>
<td></td>
</tr>
<tr>
<td>If yes, at what age did he/she learn to ride?</td>
<td></td>
</tr>
<tr>
<td>How long did he/she need training wheels?</td>
<td></td>
</tr>
<tr>
<td>Does he/she know how to swim?</td>
<td></td>
</tr>
<tr>
<td>Does he/she have problems seeing stripes on the bottom of the pool?</td>
<td></td>
</tr>
<tr>
<td>Other Health History:</td>
<td></td>
</tr>
<tr>
<td>Did the mother have pregnancy problems?</td>
<td></td>
</tr>
<tr>
<td>If yes, please explain:</td>
<td></td>
</tr>
<tr>
<td>Problems during delivery?</td>
<td></td>
</tr>
<tr>
<td>If yes, please explain:</td>
<td></td>
</tr>
<tr>
<td>Newborn: birth weight: ________           birth length: ________           head size: ________</td>
<td></td>
</tr>
<tr>
<td>Question</td>
<td>Yes</td>
</tr>
<tr>
<td>------------------------------------------------------------------------</td>
<td>-----</td>
</tr>
<tr>
<td>Was your child born prematurely (too early)?</td>
<td></td>
</tr>
<tr>
<td>If yes, how many weeks early?</td>
<td></td>
</tr>
<tr>
<td>Did the baby have any problems like:</td>
<td></td>
</tr>
<tr>
<td>☐ yellow jaundice</td>
<td></td>
</tr>
<tr>
<td>☐ meningitis</td>
<td></td>
</tr>
<tr>
<td>☐ breathing difficulties</td>
<td></td>
</tr>
<tr>
<td>☐ other:________________</td>
<td></td>
</tr>
<tr>
<td>How long did the baby stay in the hospital after birth? _______ days</td>
<td></td>
</tr>
<tr>
<td>Has your child ever been in the hospital?</td>
<td></td>
</tr>
<tr>
<td>If yes, please describe:</td>
<td></td>
</tr>
<tr>
<td>Date/age:</td>
<td></td>
</tr>
<tr>
<td>Illness or injury:</td>
<td></td>
</tr>
<tr>
<td>Does soap, lotion, medicine, etc. make your child's skin red or swollen?</td>
<td></td>
</tr>
<tr>
<td>If yes, please describe:</td>
<td></td>
</tr>
<tr>
<td>Has your child ever been treated for seizures?</td>
<td></td>
</tr>
<tr>
<td>Has your child ever been treated for fainting spells?</td>
<td></td>
</tr>
<tr>
<td>List any medication he/she takes regularly:</td>
<td></td>
</tr>
<tr>
<td>Does your child have any of the following:</td>
<td></td>
</tr>
<tr>
<td>☐ white patch of hair</td>
<td></td>
</tr>
<tr>
<td>☐ white patches of skin</td>
<td></td>
</tr>
<tr>
<td>☐ goiter or lump on neck</td>
<td></td>
</tr>
<tr>
<td>☐ cleft lip/cleft palate</td>
<td></td>
</tr>
<tr>
<td>☐ kidney problems</td>
<td></td>
</tr>
<tr>
<td>☐ diabetes</td>
<td></td>
</tr>
<tr>
<td>☐ lip pits or dimples</td>
<td></td>
</tr>
<tr>
<td>☐ extra fingers or toes</td>
<td></td>
</tr>
<tr>
<td>☐ arthritis</td>
<td></td>
</tr>
<tr>
<td>☐ bones break easily</td>
<td></td>
</tr>
<tr>
<td>☐ different color eyes</td>
<td></td>
</tr>
<tr>
<td>☐ heart murmer/defect</td>
<td></td>
</tr>
</tbody>
</table>
Questions about the Family:

This question area is about your child’s blood relatives, not relatives by marriage or adoption. The word “relation” in the tables below means how the person is related to your child, not to you. When you answer the questions, please consider both the mother’s and father’s sides of the family, including the child’s brothers, sisters, aunts, uncles, grandparents, great-grandparents, and cousins.

Please print clearly.

Ethnic background (optional; check all that apply):

- European (what country?): __________________________
- Hispanic
- American Indian
- Other (what country?): __________________________
- Asian (what country?): __________________________
- African American
- Pacific Islander

Does anyone in the family wear thick glasses, have problems seeing at night, or have any other eye problems?

<table>
<thead>
<tr>
<th>Relation</th>
<th>Eye problems:</th>
<th>Age when problem started:</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
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</tbody>
</table>

Does anyone in the family have a hearing problem (deaf, hard-of-hearing, sudden hearing loss, slow/low of hearing)?

<table>
<thead>
<tr>
<th>Relation</th>
<th>Hearing loss:</th>
<th>Age when problem started:</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
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<td></td>
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</tbody>
</table>
Does anyone in the family have neurological problems, like seizures, dizziness, fainting spells, cerebral palsy, brain damage, or any other problems with the brain, spinal cord, or nerves?

<table>
<thead>
<tr>
<th>Relation:</th>
<th>Type of neurological problems:</th>
<th>Age when problem started:</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
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<tr>
<td></td>
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</tr>
</tbody>
</table>

Who in the family does your child look like? __________________________________________

Has your child ever seen a geneticist (a doctor who knows about inheritance or how problems are passed down through families)? □ Yes □ No

If yes, who was the geneticist and where (please include address if you have that information):
________________________________________
________________________________________
________________________________________

Is there anything else we should know about your child? ________________________________
________________________________________
________________________________________
________________________________________
Student Questionnaire

Student’s name: __________________________________________________________

Date of birth: _________________________   Today’s date: ______________________

Note to interviewer:
This is to be completed for students from the age of eight or the beginning of the third grade through high school. Please take a minute to complete this questionnaire with your student or with the student and his or her interpreter.

Students with Usher syndrome may notice subtle changes associated with a gradual vision loss. Please ask the student to answer the following questions:

1. When you walk inside a dark movie theater or room, how long does it take for your eyes to adjust?
   - A couple of seconds
   - One minute
   - Longer than a minute

2. When you come inside from a bright sunny day, is it hard to see people or obstacles?
   - Yes, usually
   - Maybe sometimes
   - No, not usually

3. Do you trip over obstacles?
   - Yes, usually
   - Maybe sometimes
   - No, not usually

4. Does sunlight hurt your eyes?
   - Yes, usually
   - Maybe sometimes
   - No, not usually

5. Do you often spill liquids when pouring them into a cup or glass?
   - Yes, usually
   - Maybe sometimes
   - No, not usually

6. Can you see the stars in the sky at night?
   - Yes, usually
   - Maybe sometimes
   - No, not usually

7. If someone waves to you from the side, do you see them when you are not looking at them?
   - Yes, usually
   - Maybe sometimes
   - No, not usually

8. Is it hard to find small objects when you drop them on the floor?
   - Yes, usually
   - Maybe sometimes
   - No, not usually

9. Do you bump into people or objects within your path?
   - Yes, usually
   - Maybe sometimes
   - No, not usually

10. Do you have trouble riding a bicycle?
    - Yes, usually
     - Maybe sometimes
     - No, not usually

11. Do you sometimes knock over a glass of water or other objects on a table at mealtime?
    - Yes, usually
     - Maybe sometimes
     - No, not usually
Visual and Motor Behavior Questionnaire

As a parent, you are in a unique position to detect symptoms of learning-related vision problems in your child. These problems are not detectable by an eye chart test and can be overlooked during a quick screening. Please take a few minutes to help identify whether your child may require referral to an optometrist, ophthalmologist, or vision specialist. It should take less than three minutes to complete.

Student’s name: __________________________________________________________

Date of birth: _________________________   Today’s date: __________________

Completed by: ____________________________________________________________

Please check the box in front of the statements that best describe the following behaviors you have observed from your child. If a particular behavior does not apply, leave it blank.

Night Blindness:

□ Can’t see when coming in from bright sunlight
□ Trips over things when light changes or light is dim
□ Stays near light in a darkened room or at night
□ Positions self so light falls on the face of a person who is speaking
□ May express a desire to enter a room before it is darkened (i.e., the movie theater)
□ Avoids conversations in a darkened area
□ May appear to stagger or lose balance after an oncoming car has passed at night
□ Has problems reading under some lights or in dimly lit areas

Visual Fields:

□ Stumbles on stairs and curbs
□ Bumps into people, tables, chairs, or other objects
□ May bump or tip over objects placed to the side at mealtimes
□ Startles easily
□ Seem to hold objects in unusual position when looking at them
□ Turns head while reading across a page
□ Uses fingers to mark place while reading
□ Unable to locate small objects that have been dropped
□ Fails to glance at another person’s hand waving from the side
□ Is quiet or may edge to one side when in a large group
□ Frequently misses or fails to understand group instructions

Glare:

□ Squints and shades eyes in bright or fluorescent lighting
□ Likes to wear sunglasses even in a building but especially in bright sunlight
□ May appear awkward when exiting a building (when faced with bright lights)
Contrast:
- □ Has difficulties reading light photocopies or worksheets
- □ Unable to see stars at night
- □ Often spills when pouring a liquid
- □ Eyes become watery and red when reading

Acuity:
- □ Holds book close to eyes or bends head in awkward manner to read
- □ Sits near blackboard

Balance:
- □ Late learning to walk (past 15 months)
- □ Is considered clumsy
- □ Loses balance easily, especially in the dark
- □ Unable to ride bicycle or needed extra time to learn

Other:
- □ Is frequently last in completing group activities
- □ Exhibits anxiety in new areas
- □ Often last to enter a room
- □ May have repetitive behavior or routines
- □ May fail to participate fully in a group associated with a new situation, especially in the dark
- □ Avoids walking or running in unfamiliar areas, especially in bright sunlit areas or dark areas
- □ Constantly appears to be visually scanning a group

Does your child wear glasses or contact lenses? □ Yes □ No

If yes, □ for reading only □ for distance only □ all the time
Cone Adaptation Test

Student’s name: __________________________________________________________

Date of birth: _________________________   Today’s date: _________________________

Completed by: __________________________________________________________________________

Procedure:
Scatter the poker chips from the Cone Adaptation Test in a jumbled heap on a dark table or carpet. The lights in the room should be dimmed to imitate twilight or dusk. The light needs to be even in the room. To check if the lights are dimmed to the correct level, ask if the student can see you signing. If the student can see you sign, the room is too bright.

Step 1: Ask the student to pick up the white chips. Observe how long it takes for him or her to complete the task. Make note of the start time, stop time, and total time.
Step 2: Then ask the student to sort the red chips from the blue chips. Make note of the start time, stop time, and total time it took to complete the task.
Step 3: Document the number of chips correctly sorted.
Step 4: Repeat steps 1–3.

Results:

<table>
<thead>
<tr>
<th>Trial #1</th>
<th></th>
<th>Trial #2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Time stopped:</td>
<td></td>
<td>Time stopped:</td>
</tr>
<tr>
<td>Total time:</td>
<td></td>
<td>Total time:</td>
</tr>
<tr>
<td>Number correct:</td>
<td></td>
<td>Number correct:</td>
</tr>
<tr>
<td>Red:</td>
<td>Blue:</td>
<td>White:</td>
</tr>
<tr>
<td>Red:</td>
<td>Blue:</td>
<td>White:</td>
</tr>
</tbody>
</table>

Mark as pass (P) if the student sorts chips correctly within two minutes. Mark as fail (F) if the student has difficulty sorting chips by color or within two minutes.

Action:

Pass: The family interview showed no family history of deafness with vision loss and no indication of nightblindness, field loss, glare sensitivity, acuity loss, or balance problems and the student passed all stations.

Retest in one year: The family interview was incomplete or family history is uncertain, the family history indicated problems in one area of concern, the student didn’t understand directions during stations, or the student had difficulty with one station.

Refer to ophthalmologist: Family interview reflects history of combined hearing and vision loss, and/or symptoms of acuity loss, nightblindness, field loss, glare sensitivity, or balance problems and the student failed one station.
Visual Field Tests

Student's name: __________________________________________________________

Date of birth: _________________________    Today’s date: _________________________

Completed by: ______________________________________________________________

Procedure: Conduct either the Wiggling Fingers Test or the Two Fingers Test.

Wiggling Fingers Test:
This test should be conducted with both of the student’s eyes open. You and the student should be seated facing each other at approximately one arm’s length. Tell the student to look at your nose and tell you when he or she sees your fingers. Extend both of your arms to the side of your body. Bend both of your wrists and begin wiggling your fingers while moving your arms closer to the student. Document at which point the student first sees your wiggling fingers. Continue bringing your hands in closer toward your body to detect ring scotomas. Conduct the test for both horizontal and vertical planes.

OR

Two Fingers Test:
This test should be conducted with both of the student’s eyes open. You and the student should be seated facing each other at approximately one arm’s length. Tell the student to look at your nose. Hold one or two fingers to the side of your own visual field. Ask the student whether one or two fingers are present. Document the point at which the student can see the fingers. Continue bringing your hands in closer toward your body to detect ring scotomas. Conduct this test for both horizontal and vertical planes.

Color in the areas the students was able to see. Use a dark color, not a highlighter. Any section of the semicircle not colored in means that the student failed the screening.

Color in the areas the student was able to see. Use a dark color, not a highlighter. Any section of the semicircle not colored in means that the student failed the screening.
## Balance Test

**Student's name:** ____________________________________________________________

**Date of birth:** _________________________    **Today’s date:** __________________________

**Completed by:** ____________________________________________________________________

**Procedure:** Conduct both of these tests.

**Feet together:**
Have the student stand with feet together, arms stretched out to the side of the body, and eyes closed. Stand behind the student while gently pushing on either side of the student’s torso. (Note: The student may lose balance quickly, so be ready to support him or her.) Document whether the student was able to maintain his/her balance. Conduct the same procedure again but this time with the student’s eyes open. Document the student’s reaction.

**Results:**
Mark pass (P) if the student can regain balance after gentle push. Mark fail (F) if the student has difficulty maintaining balance even without push, or cannot regain balance after push.

<table>
<thead>
<tr>
<th>Balance maintained</th>
<th>Balance not maintained</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>eyes closed, feet together</td>
</tr>
<tr>
<td></td>
<td>eyes open, feet together</td>
</tr>
</tbody>
</table>

**One Foot in Front of the Other:**
Have the student stand with one foot in front of the other, arms stretched out to the side of the body, and eyes closed. Stand behind the student while gently pushing on either side of the student’s torso. (Note: The student may lose balance quickly, so be ready to support him or her.) Document whether the student was able to maintain his/her balance. Conduct the same procedure again but this time with the student’s eyes open. Document the student’s reaction.

**Results:**
Mark pass (P) if the student can regain balance after gentle push. Mark fail (F) if the student has difficulty maintaining balance even without push, or cannot regain balance after push.

<table>
<thead>
<tr>
<th>Balance maintained</th>
<th>Balance not maintained</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>eyes closed, one foot in front of the other</td>
</tr>
<tr>
<td></td>
<td>eyes open, one food in front of the other</td>
</tr>
</tbody>
</table>
# Usher Screening Data Summary

Student’s name: ____________________________________________________________

School district: ___________________________    School: ___________________________

Date of birth: ___________________________    Today’s date: ___________________________

Completed by: ____________________________________________________________

<table>
<thead>
<tr>
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</thead>
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<tr>
<td>Family History Questionnaire</td>
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</tr>
<tr>
<td>Student Questionnaire</td>
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### Visual and Motor Behavior Questionnaire Forms

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</tr>
</thead>
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</tr>
<tr>
<td>Teacher</td>
<td>□ Yes □ No</td>
</tr>
<tr>
<td>Other</td>
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</tbody>
</table>

### Behaviors

<table>
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</thead>
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<td>Nightblindness</td>
<td>□ Yes □ No □ Yes □ No</td>
</tr>
<tr>
<td>Visual Fields</td>
<td>□ Yes □ No □ Yes □ No</td>
</tr>
<tr>
<td>Glare</td>
<td>□ Yes □ No □ Yes □ No</td>
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<tr>
<td>Contrast</td>
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</tr>
<tr>
<td>Acuity</td>
<td>□ Yes □ No □ Yes □ No</td>
</tr>
<tr>
<td>Balance</td>
<td>□ Yes □ No □ Yes □ No</td>
</tr>
<tr>
<td>Other</td>
<td>□ Yes □ No □ Yes □ No</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Completed?</th>
<th>Appears Normal?</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cone Adaptation</td>
<td>□ Yes □ No □ Yes □ No</td>
</tr>
<tr>
<td>Field Screening</td>
<td>□ Yes □ No □ Yes □ No</td>
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<tr>
<td>Balance Screening</td>
<td>□ Yes □ No □ Yes □ No</td>
</tr>
<tr>
<td>Section</td>
<td>Recommend Setting?</td>
</tr>
<tr>
<td>-----------------</td>
<td>--------------------</td>
</tr>
<tr>
<td>Referral Letter</td>
<td>☐ Yes   ☐ No</td>
</tr>
<tr>
<td>Follow Up</td>
<td>☐ Yes   ☐ No</td>
</tr>
</tbody>
</table>

Comments:
Appendix F: Glossary

**acuity loss**: related to nearsightedness (the ability to see objects close to the body, but not at a distance) or farsightedness (the ability to see objects at a distance, but not close to the body).

**ASL**: an acronym for American Sign Language.

**assistive listening devices**: units that are equipped with a microphone and receiver that helps the individual listen to the speaker while interrupting surrounding conversations and environmental sounds.

**audiogram**: a chart that indicates what a person does and does not hear in response to the speed and loudness of sound.

**bilateral hearing loss**: a hearing loss in both ears.

**blind spots**: spots that affect the visual field. They may occur as a spot, a series of spots, or in a ring. Blind spots also are referred to as scotomas.

**blindness**: an impairment in vision in which the person has 20/200 with correction and/or who has a 20 degree visual field is considered legally blind.

**central acuity loss**: a vision loss that occurs in the central part of the retina that reduces the person’s central field of vision, acuity, ability to see detail, and color vision.

**CHARGE syndrome**: a genetic condition, resulting in colobomas of the eye, heart defects, nasal atresia, delayed growth, genital involvement in males, and ear involvement including deafness.

**conductive hearing loss**: a loss that occurs in the middle ear usually related to infection or malfunction of the structures in the middle ear. This loss typically can be corrected with treatment.

**cones**: the part of the retina that allows the eye to see color.

**congenital deafness**: a person who is born deaf or hard-of-hearing.

**deaf-blindness**: concomitant hearing and visual impairments, the combination of which causes such severe communication and other developmental and educational needs that they cannot be accommodated in special education programs solely for children with deafness or blindness.

**deafness**: an impairment in hearing so severe that the child is impaired in processing linguistic information through hearing, with or without amplification, which adversely affects development or educational performance.

**decibels**: the measure of intensity or loudness of sound. The notation for decibels is dB.
electroretinogram (ERG): a test conducted by an ophthalmologist that measures the electricity discharged by the nerve impulses in the retina. (For more about ERG, see Appendix A: What Is an Electroretinogram (ERG) Evaluation?)

geneticist: a doctor who detects genetic conditions and provides genetic counseling.

Goldenhaar syndrome: an inherited condition resulting in irregular facial structure, skin folds located on the lower corner of the eyeball, astigmatism, deafness, and spinal column involvement.

hard of hearing: a hearing loss where the person is able to hear or detect some sounds at various decibels and hertz.

hearing impairment: an impairment in hearing that is so severe the child is impaired in processing linguistic information through hearing, with or without amplification, which adversely affects development or educational performance.

hertz: the frequency or rate at which sounds travel. The notation for hertz is hz.

OD: a notation in a doctor’s report that indicates right eye.

ophthalmologist: a medical doctor who assesses and diagnoses visual conditions and performs medical interventions based on the visual conditions.

OS: a notation in a doctor’s report that indicates left eye.

OU: a notation in a doctor’s report that indicates both eyes.

Refsum syndrome: an inherited condition characterized by changes in metabolism resulting in deafness, vision loss, and balance difficulties, which are degenerative.

retina: the structure that contains the rods and cones of the eye that allows color vision, detail vision, night vision, and the ability to see darkness affecting the retina. It also affects the blood vessels.

retinitis pigmentosa (RP): a progressive, genetic condition that reduces the peripheral field of vision and the ability to see darkness affecting the retina. It also affects the blood vessels.

rods: the part of the retina that allows the eye to see movement and in the dark.

rubella: a viral infection that is transmitted to the fetus during pregnancy resulting in cataracts, glaucoma, myopia, hearing loss, heart disease, and intellectual disability. The severity of these conditions is dependent upon the time it is acquired.

scotoma: a blind spot or spots in the visual field.

SEE: a sign language system that stands for Signing Exact English.
**sensorineural hearing loss**: a hearing loss that occurs in the inner ear usually related to malfunction of the structures in the inner ear. The loss typically cannot be corrected with treatment.

**unilateral hearing loss**: a hearing loss in one ear.

**visual impairment**: a visual impairment that, even with correction, adversely affects a child’s development or educational performance. The term includes both partially sighted and blind.
Appendix G: Resources

State and Regional Resources

Conklin Center for the Blind – http://www.conklincenter.org/
Deaf Service Bureau – http://www.deafservicebureau.org/
Deaf Service Center Association – http://www.fldsca.org/
Florida Association for Education and Rehabilitation of the Blind and Visually Impaired – http://www.flaer.org/
Florida Deaf-Blind Association – http://fldeafblind.org/
Florida Division of Blind Services – http://dbs.myflorida.com/
Florida Instructional Materials Center for the Visually Impaired – http://www.fimcvi.org/
Florida Registry of Interpreters for the Deaf – http://www.fridcentral.com/
Florida Outreach Project for Children and Young Adults Who Are Deaf-Blind – http://www.deafblind.ufl.edu/index.htm
Florida School for the Deaf and the Blind – http://www.fsdb.k12.fl.us/
Hands and Voices – http://handsandvoices.org/
Helping the Blind and Visually Impaired – http://visuallyimpairedandtheblind.com/index.html

Lighthouses for the Blind:

  Big Bend – http://www.firesight.org/
  Broward – http://www.lhob.org/
  Central Florida – https://www.lighthousecentralflorida.org/
  Miami – http://www.miamilighthouse.com/
  Palm Beaches – http://www.lighthousepalmbeaches.org/
  Pinellas – http://www.lighthouseofpinellas.org/
  Tampa – http://www.tampalighthouse.org/
  West Pasco and Hernando – http://lvib.org/
National Council of Private Agencies Serving the Blind and Visually Impaired –
http://www.agenciesfortheblind.org/ByLocation.asp#Florida


fl.us/

National Resource Agencies


org/Pages/default.aspx

The Foundation for Fighting Blindness (National Retinitis Pigmentosa Foundation) –
http://www.blindness.org/

Helen Keller National Center – http://www.hknc.org/

National Consortium on Deaf-Blindness – http://www.nationaldb.org/

National Information for the Center on Deafness – http://www.nidcd.nih.gov/
References


