

Screening for Usher Syndrome

A hands-on guide for school nurses

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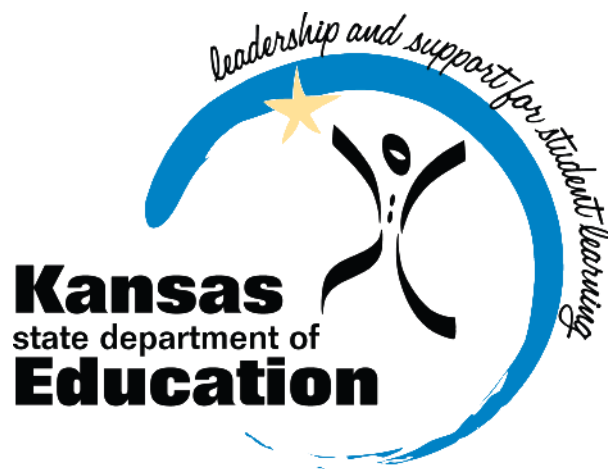
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The members of the Kansas Usher Screening Committee

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Preface

This manual was written specifically to help school nurses conduct screenings for Usher syndrome, a genetic condition that involves deafness or hearing loss and the progressive loss of vision. As such, it provides the step-by-step process of how to conduct a screening, the actual forms needed for a screening, and resources for referring students for further evaluations and counseling.

Please be aware that members of the Kansas Usher Screening Committee are available to help conduct Usher syndrome screenings. To request on-site assistance, nurses should contact the Director of the Kansas Deaf-Blind Project, Kansas State School for the Blind, 1100 State Avenue, Kansas City, KS 66102-4411, Phone: 913-305-3014.

The Kansas Usher Screening Committee was formed in 1996 in response to a growing national awareness of the importance of screening for Usher syndrome. The committee began meeting in 1996 to learn how to conduct educational screenings for students who may have Usher syndrome. The committee began meeting in 1996 to learn how to conduct educational screenings for students who may have Usher syndrome. Committee members met for a year under the guidance of Dr. Sandra Davenport, a leading national expert who pioneered educational screening efforts for Usher syndrome. Two pilot site screenings were conducted: one at Kansas State School for the Deaf and one in Wichita Public Schools. A total of 79 students participated in the screenings. Of this total, 37 students passed both paper and site screenings, seven students were referred for an ERG, 25 students were targeted for rechecks within one year, and 10 students were targeted for vision evaluations other than an ERG. One of the two students who were referred for an ERG was diagnosed with Usher syndrome.

A collaborative effort between the Kansas Department of Health and Environment and the Kansas State Department of Education was initiated to include testing for Usher syndrome with the vision screening certificate training offered every year at various locations throughout the state. The training combined with the development of resource packages on Usher syndrome and this procedural manual should help increase the awareness and implementation of screening programs. Through this collaboration from members of the Kansas Usher Screening Committee, students who have this syndrome and their families will receive appropriate educational services and supports.

Section I: Introduction

What is Usher Syndrome?

Usher syndrome is a genetic condition that involves deafness or hearing loss and the progressive loss of vision due to Retinitis Pigmentosa (RP) (For more information on Retinitis Pigmentosa, see “What is Retinitis Pigmentosa?,” page 22). In addition, depending on the type of Usher syndrome acquired, it may affect a person’s sense of balance.

There are three types of Usher syndrome. Different characteristics of vision, hearing, and balance are associated with each type. (For a list of the different characteristics associated with each type of Usher syndrome and a description of the different types of hearing loss, see page 23.)

A person with Usher syndrome Type I typically is born with a profound hearing loss in both ears, which is characterized by a corner audiogram. (See Diagram 1 for an illustration of a corner audiogram).A visual field loss usually is detected in early elementary years and progresses to total blindness as an adult. Finally, a person’s sense of balance is affected.

Diagram 1: Corner Audiogram

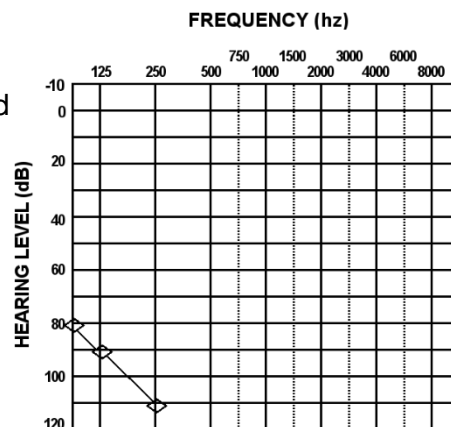
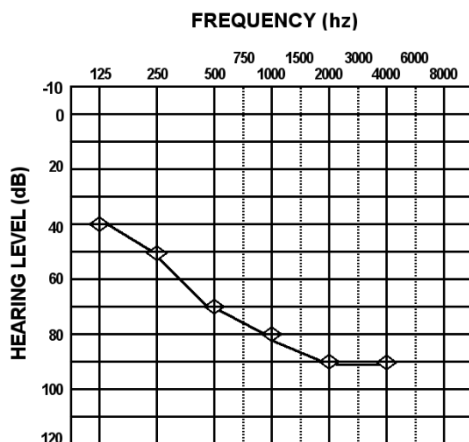


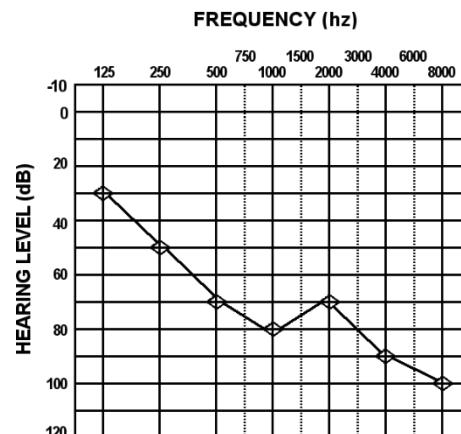
Diagram 2: Sloping Audiogram



A person with Usher syndrome Type II typically is 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.) Vision loss usually occurs during the mid to late teenage years and does not usually progress to total blindness. The person’s balance is not affected.

A person with Usher syndrome Type III (the most rare) typically is born with normal hearing or a mild to moderate hearing loss in both ears: however, the hearing loss may drop drastically within a five to ten year period. This loss is characterized by an audiogram with a slope and a ski bump. (See Diagram 3 for an illustration of this type of audiogram.) Vision loss progresses the same as Usher syndrome Type II; however, legal blindness may occur from 20 to 40 years of age. Some individuals may experience balance problems.

Diagram 3: Ski Bump Audiogram



How Does a Person Inherit Usher Syndrome?

Usher syndrome is an “autosomal recessive condition.” “Autosomal” means that the genes are not sex linked. Males and females are affected equally. “Recessive” means that the Usher gene is inherited from each parent. Most parents do not know they are a carrier of Usher syndrome until their child is diagnosed. The possible combinations that will result in Usher syndrome are:

- If both parents are carriers, the chances are one in four children will have Usher syndrome.
- If only one parent has Usher syndrome and the other is not a carrier, no children will be affected; however, all will be carriers.
- If one parent has Usher syndrome and the other parent is a carrier, two of four children (50%) will have Usher syndrome.
- If both parents have Usher syndrome, all of the children will have Usher syndrome.

Why is Screening Important?

Screening for Usher syndrome is important because students with Usher syndrome usually require specific accommodations and modifications in order to function effectively and safely. They also have behaviors that are often misinterpreted as clumsy, slow, and uncooperative.

Thus, screening may identify students who need to learn how to safely function within different environments, students who need additional related services in school (e.g., career planning, genetic counseling, and social or emotional counseling), and students who need modifications and adaptations during instruction. Once Usher syndrome is diagnosed, students can better prepare for the changes facing them as the condition progresses.

Caution: Screening can only *detect* early signs of Usher syndrome. For an accurate diagnosis, an ophthalmological evaluation if an electroretinogram (ERG) and extensive visual field testing are required. (For more information on ERGs, see “What is an Electroretinogram Evaluation?” page 24.) Additionally, genetic counseling is recommended to confirm the student has Usher syndrome.

How is the Screening Conducted?

Basically, there are two parts to a screening: the paper screening and the station screening. The purpose of the paper screening is twofold: a) to target candidates for the station screening, and b) to eliminate students from participation in further screenings. The paper screening identifies family history that may indicate the potential for Usher syndrome and behavior characteristics that may indicate a visual field loss, blind spots, or acuity loss. (Refer to page 4 to learn how to conduct a paper screening.)

The station screening consists of three stations: visual field, cone adaptation, and balance. The visual field and cone adaptation stations are used to identify students who demonstrate visual behaviors that are characteristic of RP, rod deterioration, or cone deterioration. The balance station is used to identify students who demonstrate balance difficulties with the lights on or the lights off. (Refer to pages 7-12 to learn how to conduct a station screening.)

Help! Where Do I Begin?

Before you begin a screening, familiarize yourself with the different types of Usher syndrome, the inheritance patterns, and why screenings are important (pages 1-3). Next, do the following:

1. Read Sections II, III and IV of this manual.
2. Copy the forms needed for the paper screening. These are listed on page 4.
3. After the paper screening has been conducted, determine if a station screening is required. (See pages 4-6 for making this determination.)
4. Obtain a Cone Adaptation Test if you do not have one. (See page 10)
5. Copy the forms necessary for the station screening. (See page 6)
6. Determine if you need help conducting the station screening. If you do, contact Director of the Kansas Deaf-Blind Project, Kansas State School for the Blind, 1100 State Avenue, Kansas City, KS 66102-4411, Phone: 913-305-3014. Remember: Members of the Usher Screening Committee are available to help.
7. Evaluate the results from the paper screening and the station screening. Determine referral status of the student by following the instructions in Section IV. If you are not sure about the results, read the case study in Appendix B.
8. Review resources in Section VII. Refer school administrators, teachers, and family members to experienced evaluation personnel located in this section.
9. Once the student has been diagnosed, give school administrators, teachers, family members and, if appropriate, the student (age 18 and his or her own guardian) A Collection of Resources on Usher Syndrome Packet that contains useful information of Usher syndrome. (Refer to Section VII to obtain a packet.)
10. Contact the Director of the Kansas Deaf-Blind Project for processing certification on the Kansas State Deaf-Blind Census.

Section II: The Paper Screening

Prior to the Paper Screening

- A. Identify students who have the characteristic hearing loss as demonstrated by audiogram (see Diagrams 1-3, page 1).
- B. Send a letter from the school to parents/guardians of these students. Include the following materials from Appendix C:
 - *Kansas Usher Screening Project Family Questionnaire*
 - *Kansas Usher Screening Project Behavior Questionnaire*
 - Your district's release of information form
 - Self-addressed stamped envelope, if possible.

Timeline:

Allow two weeks from the date of the parent/guardian letter for families to return materials. If materials are not returned within this time, contact families by telephone to encourage their involvement in the screening process.

Upon Receipt of the Family Information

- A. Obtain the following completed information:
 - Kansas Usher Screening Project Behavior Questionnaire (from the student's teachers and dorm staff, if appropriate)
 - Kansas Usher Screening Project Student Questionnaire
Note: the Student Questionnaire should be completed for students from 3rd grade through 12th grade. The student's teacher should interview the student to complete the form.

Timeline:

Allow one month to obtain the above questionnaires.

After You Have Received All Materials

- A. Review all materials and determine if the student should receive a station screening. Students who meet at least HALF of the criteria on each of the following items should receive a station screening.

Item #1: Family History Questionnaire

- The student has not been diagnosed with another syndrome (e.g., Goldenhar or Rubella).
- The student did not have multiple organs effected at the time of birth or as a result of a degenerative condition.
- The student's family history indicates no multiple generations of deafness or hard of hearing, or neither biological parent is deaf or hard of hearing.
- There is no incidence of mental retardation.
- The student walked earlier than 18-24 months.
- The student was born deaf or hard of hearing (bilateral sensorineural loss). Or, the student was born with normal hearing and it dropped significantly within a 5-10 year period and that drop 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) 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 two relatives from past generations who were born deaf or hard of hearing and lost their sight in their midteens to early 30s, the student should receive a station screening. If both biological parents have Usher syndrome, the student definitely needs to receive a station screening.

Item #2: Audiogram

- The student's audiogram shows no gradual changes over time.
- The shape of the student's audiogram is sloped or dropped.
- The slope of the loss depicted on the student's audiogram does not rise in the higher frequencies.
- The audiogram only shows one ear with a unilateral, sensorineural loss.

Note: A corner audiogram with losses in the severe to profound range and lower frequencies, a sloping audiogram, or a sloping audiogram with a ski bump would indicate a need for a station screening.

Item #3: Student Behavior Questionnaire

- 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 is night blind.
- 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.
- The student is unable to function in low contrast situations.

Note: If more than one person marks the same behaviors on each section of the form, this may indicate a visual field loss or balance difficulties. The student should be referred to a station screening.

Item #4: Student Interview Questionnaire (if applicable)

- 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.

Note: A large number of matching responses (e.g. half or more) on the Student Interview Questionnaire and the Student Behavior Questionnaire may indicate visual field losses or balance difficulty. The student should receive a station screening.

B. Develop pocket folder for each student to be screened with the following information in each folder:

Completed Forms:

- Student audiograms (current and previous)
- Family Questionnaire
- Behavior Questionnaires (teacher(s), parent, and dorm teacher if student lives in residential setting)
- Release of Information

Blank Forms:

- Cone Adaptation Form
- Balance Screening Form
- Field Screening Form
- Kansas Usher and Vision Screening Data Summary Form

Section III: The Station Screening

Preparation

- A. 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.
- B. Plan for the screening day. This includes:
 1. Gather the following materials:
 - Cone Adaptation Test
 - Light meter
 - 2 timers or stop watches
 - Students' pocket folders
 2. Arrange for interpreters to match communication needs of the students (e.g., ASL, SEE, or CASE).
 3. Identify rooms for each station (i.e., cone, field, and balance) and determine if appropriate:
 - a. Cone: Can the room be made dark enough for testing?
 - b. Field: It should be a small room with good lighting and three chairs (evaluator, interpreter, and student).
 - c. Balance: It should be a room large enough for two evaluators, two interpreters, and two students. Each student should be able to extend both arms out to the side of his or her body.
 4. Recruit people to assist with the screening (Contact the Director of the Kansas Deaf-Blind Project for help in identifying possible screeners).
 5. Develop a schedule for screening (e.g., screen elementary, middle school, high school students separately; groups should consist of no more than four students at one time).
 6. Inform teachers of time, day, and location of the screening in order to release students from class.
 7. Hold a planning meeting with all screeners to run through the screening process. (See example of Usher Screening Inservice Agenda, page 25.)
 - a. Identify who will work at each station.
 - b. Identify someone to assign students to stations and give them folders when they first enter the screening area.
 - c. Explain how to complete the forms.
 - d. Have screeners practice the process on each other.
 - e. Complete any of the tasks that require a great deal of time (e.g., review completed questionnaires to determine which students may have difficulties during the station screening).

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. To avoid burdening school nursing staff, screeners can be recruited from within the district or from the Kansas Usher Screening Committee. Examples of possible district staff include consulting or on-staff speech therapists, audiologists, teachers of vision or hearing, and medical doctors or nurses in the neighboring community. For assistance from the Kansas Usher Screening Committee, contact the Director of the Kansas Deaf-Blind Project, Kansas State School for the Blind, 1100 State Avenue, Kansas City, KS 66102-4411, Phone: 913-305-3014).

The total number of screeners needed will depend upon the number of stations and number of students needing the station screening. If only a handful of students require the station screening, only two or three screeners will be needed. However, if more than five students will receive the station screening, at least one screener should be identified for each station.

To insure consistency and accuracy of test results, it is essential that screeners meet in advance of the station screening to learn about Usher syndrome, understand the purpose of screening, and develop clear expectations for the screening process by practicing the various tests on each other. A sample in-service agenda is included on page 25 to facilitate preparation for the station screening day.

The Role of the Interpreter

In order for the screening process to be valid, interpreters must be provided for students who rely on a visual, tactual, or oral interpreter in the classroom. In addition, interpreters must match the communication needs of the student (e.g., students utilizing American Sign Language (ASL) paired with ASL interpreters; students utilizing 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 (ALD), glasses or other visual aids, these must be used during the screening process.

The total number of interpreters needed for the screening will depend upon the number of screeners at each station. The number of students being screened and the room layout will determine the number of screeners and interpreters. The approximate number of interpreters needed for each activity is:

Activity	Number of Interpreters
Welcome/Explanation	1
Cone Adaptation	1-2
Balance	2
Field Screening	1
TOTAL	5-6

Interpreters should be assigned to one station and remain there during the duration of the screening process. This will minimize confusion and ensure consistency. 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. For tactile interpreting, breaks should be provided every 15 to 30 minutes.

A. The Screening

Welcome students and explain procedures

People and Materials Needed:

- Greeter (greet students, gives them their folder, and directs them to stations)
- Explainer (someone familiar with the screening process)
- Mock student (another staff person to role-play)
- Interpreter (using communication skill of the students)
- Cone Adaptation Test

Room Setup:

Select a room large enough to accommodate an explainer, an interpreter, a mock student, and up to four students. During this activity, students may choose to stand or be seated. However, students must be able to clearly see the role play and the interpreter.

Procedures:

The greeter welcomes the students (there should be no more than four students in each group) and hands each of them their student folder. The greeter then directs all of the students into the room for the explanation and demonstration. The explainer briefly describes Usher syndrome and explains the screening process to the students. (See “How to Explain Usher Syndrome to Kids,” page 26.) The explainer and mock student then demonstrate each screening activity (i.e., Cone Adaptation Test, the Balance Test, and the Field Screening). Care should be taken to ensure that the demonstration and instruction at this station match the demonstration and instruction at the other stations. Students should be asked if they have any questions, and answers provided accordingly. Once all questions are answered, the greeter should direct students to the different stations: one to the Field Screening Station, two to the Balance Screening Station, and one to Cone Adaptation Station.

Timeline:

15-20 minutes or as long as necessary to answer questions

B. Station 1: Cone Adaptation Test

People and Materials Needed:

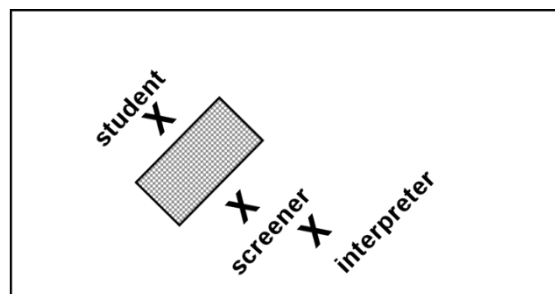
- Screener (may utilize two screeners if two separate rooms with the same light level are available)
- Interpreters (equal to the number of screeners)

For each screening room:

- Cone Adaptation Test
- Night light
- Table with dark top
- Three chairs
- Two timers or stop watches
- 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 possible 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. (See diagram at right.)



Procedures to be followed by screener:

- a) Review procedures on Cone Adaptation Form for more detailed instructions.
- b) Re-explain the Cone Adaptation Test.
- c) Explain that when the light is turned off, the student begins the activity.
- d) Conduct the Cone Adaptation Test and document results on the student's Cone Adaptation Form.
- e) Reinforce the student for completing the test.
- f) Conduct the Cone Adaptation Test and document results again.
- g) Reinforce the student for completing the second test. The screener may choose to share the results with the student.
- h) Return Cone Adaptation Form to the student folder. Give the folder to the student.

Notes:

1. Document on the screening form any irregularities that occur (e.g., squares fall on the floor; the student uses his or her hands instead of both eyes to locate spares).
2. For young children (preschoolers through third grade), explain the activity, give the student an opportunity to practice sorting and stacking the squares, and then conduct the test with the lights off.

Timeline:

Allow 15 minutes per student

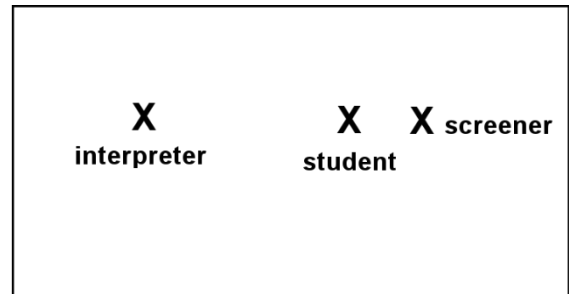
C. Station 2: Balance Test

People and Materials Needed:

- Two screeners
- Two interpreters
- Balance Form (within student folder)

Room Setup:

Unlike Cone Adaptation Test, two students can be screened for Balance in the same room. Each student should stand facing an Interpreter. One screener should stand behind each student (See Diagram at right.)



Procedures to be followed by screener:

- Review procedures on the Balance Form for detailed instructions.
- Re-explain the procedures for the Balance Test.
- Tell the student to see if he can keep his balance during the first component of the test.
- Conduct the first component of the Balance Test.
- 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.
- Conduct the second component of the Balance Test.
- Reinforce student for completing the test.
- Document results.
- Return Balance Form to student folder.

Timeline:

Allow 10 minutes per student

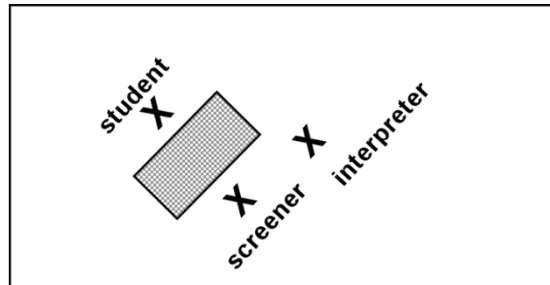
D. Station 3: Field Screening

People and Material Needed:

- One screener
- One interpreter
- Three chairs
- Field Screening Form (within student folder)

Room Setup:

Both the screener and student should be seated facing each other. There should be approximately one arm's length between them. The interpreter should be standing behind the screener. (See diagram at right.)



Procedures to be followed by screener:

- a) Review procedures on the Visual Field Form for more detailed instructions.
- b) Re-explain the procedures for the Field Screening.
- c) Determine which test you will conduct.
- d) Explain the activity to the student.
- e) Conduct the activity.
- f) Reinforce the student for completing the test.
- g) Document results.
- h) Return the Field Screening Form to the student folder.

Note:

If the student has only one eye, make note on the results of the Field Screening Form.

Timeline:

Allow 10 minutes to complete the activity.

Section IV: Evaluating the Results

Once you have completed the paper and station screening, compile data for each student onto an *Usher Screening Data Summary Sheet* (Appendix C). Obtain missing data before proceeding.

Once the form has been completed, there are four possible actions you can take: a) pass the student (i.e., eliminate from further screenings), b) re-check the student within one year, c) refer the student for an ophthalmological evaluation, or d) refer the student for an ERG and visual field testing. The criteria for each of these actions are summarized below.

Pass the student if:

1. Neither biological parent is deaf OR:
 - the family history does not show a history of deafness with a visual impairment,
 - the student began walking at the normal development age, OR
 - the student does not show difficulty with balance.
2. The student has been diagnosed with a syndrome other than Usher syndrome or multiple organs were affected at the time of birth that were not the result of trauma, disease, or infection.
3. The student's audiogram does not reflect a corner, a slope, or a slope with a ski bump.
4. The student wears glasses and the family history, student interview, and the behavior checklists do not reflect a pattern of visual field loss.
5. The student passed all three station screenings (cone adaptation, balance, and visual field), OR:
 - The student passed the Visual Field Test, passed 75% or better of the Cone Adaptation Test (within a two-minute time period), and passed 75% or better of the Balance Test, OR
 - The student passed the Visual Field Test and Cone Adaptation Test, and 75% of the Balance Test.

If the student meets all of the above criteria, the results indicate a pass. If the family history shows a inconsistent pattern of responses and the behavior checklist indicates possible symptoms of a visual field loss, re-check the student in one year (with both paper and station screenings).

Re-check the student in one year if:

1. The student is under the age of six or had difficulty understanding the testing instructions or demonstration.
2. The family history is unclear as to a past history of hearing loss with a visual impairment, the student did not begin walking at the normal developmental age, or the student has difficulty with balance.
3. The student has multiple organs that were affected after birth due to infection, disease, or trauma.
4. The student's audiogram reflects a corner, a slope, or a slope with a ski bump.
5. 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.
6. The student passed all three station screenings (cone adaptation, balance, and visual field), OR the student passed the Visual Field test, less than 75% of the Cone Adaptation Test (within a two-minute time period), and less than 75% of the Balance Test.

If the student meets all of the above criteria, the results indicate a re-check. If the family history shows an inconsistent pattern of responses and the behavior checklist indicates possible symptoms of a visual field loss, refer the student for an ERG. (It is better to refer on the side of caution.)

Refer for an ophthalmological evaluation if:

1. The family history does not indicate a history of hearing loss with a visual impairment, the student began walking at the normal developmental age, and the student has difficulty with balance.
2. The student does not have multiple organs affected or the organs were affected at the time of birth.
3. The student's audiogram reflects a corner, a slope, or a slope with a ski bump, OR the audiogram shows a consistent pattern of loss over time.
4. The family history, student interview, and behavior checklists reflect symptoms of acuity loss, blind spots, eye muscle imbalance, glare and contrast sensitivity, or color blindness.
5. The student passed all three station screenings (visual field, cone adaptation, and balance) OR the student passed the Visual Field Test, more than 75% of the Cone Adaptation Test (within a two-minute time period), and less than 75% of the Balance Test.

If the student meets all of the above criteria, the results of the test indicate a need for an ophthalmologic evaluation other than an ERG.

Refer for an ERG and extensive visual field evaluation if:

1. The family history indicates both biological parents have Usher syndrome, one parent has Usher syndrome, or a past history of hearing losses with a visual impairment.
2. The student did not begin walking within the normal developmental age and has difficulty with balance.
3. The student does not or does have multiple organs affected or the organs were affected after birth due to trauma, illness, or disease.
4. The student's audiogram reflects a corner, a slope, or a slope with a ski bump.
5. The student behaviors from the family history, student interview, and behavior checklists reflects symptoms of visual field loss and night blindness.
6. The student did not pass the Visual Field Test or was not able to test.
7. The student passed less than 75% of the Cone Adaptation Test (within a two-minute time period).
8. The student passed less than 75% of the Balance Test.

If the student meets all of the above criteria, refer for an ERG and extensive visual field evaluation.

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Section VI: 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: a sign language system that stands for America 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 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: a person who sees 20/200 with correction and/or who has a 20 degree visual field.

CASE: a sign language system that stands for Conceptually Accurate Signed English.

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 Association: a condition in which the cause is unknown, 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.

deafness: not able to hear or detect the production of sounds.

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 "What is an Electroretinogram?", page 24.)

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

Goldenhar 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.

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

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

low vision: a person who sees approximately 20/60-20/70 to 20/200 with correction or who has 40 degree visual field. Sometimes referred to as partially sighted.

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. (For more information, see "What is Retinitis Pigmentosa?", page 22.)

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 mental retardation. 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.

Section VII: Resources

WRITTEN & VISUAL RESOURCES

Balkwill, F., & Ralph, M. (1995). *How did I get Usher syndrome?* London: Harper Collins Publishers. Information on how Usher syndrome is acquired as presented in a comic book format with an eyeball serving as narrator. The eye takes a young woman through a journey of how she inherited Usher syndrome, and the impact that Usher syndrome has on her activities of daily living.

National Deaf-Blind & Rubella Association
11-13 Clifton Terrace
Finsbury Park, London N4 3SR
Phone: 0171 272 7774

Davenport, S.L.H. (1996). *Important topics in deafblind education; Usher syndrome I, II, and III.* (videotape). Terre Haute: Blumberg Center for Interdisciplinary Studies in Education, Indiana State University. This 20-minute tape presents the characteristics inherent in all three types of Usher syndrome. Typically, students who have Usher syndrome I will experience visual field loss early, are born profoundly deaf, and have poor balance. Students with Usher syndrome II begin losing vision during their late teens, are born with a moderate to profound hearing loss, but do not experience balance problems. Students with Usher syndrome III are very rare. These students have a visual field loss similar to those with Usher syndrome II, are born with normal hearing or have a mild hearing loss that rapidly decreases within a period of 10 years, and may or may not have balance problems. Some preliminary characteristics and suggested modifications to compensate for the vision loss and balance difficulties are described.

Kansas Deaf-Blind Loan Library
Kansas Instructional Resource Center
Kansas School for the Blind, 1100 State Ave.
Kansas City, Kansas 66102
1-800-572-5463, ext. 5

Davenport, S.L.H. (1996). *Important topics in deaf-blind education: Patterns of inheritance.* (videotape). Terre Haute: Blumberg Center for Interdisciplinary Studies in Education, Indiana State University. This 12-minute videotape presents the basics of genetics and traits inherited by males and females. Recessive, dominant, and X-linked characteristics are passed through generations. These traits have significant impact on the physical body of individuals in various forms. Several conditions are presented and discussed, such as Tay Sachs, Stickler syndrome, Treacher Collins syndrome, PKU, and Usher. Many of these conditions affect both vision and hearing sensory systems

Kansas Deaf-Blind Loan Library
Kansas Instructional Resource Center
Kansas School for the Blind, 1100 State Ave.
Kansas City, Kansas 66102
1-800-572-5463, ext. 5

Davenport, S.L.H. (1997). *Important topics in deafblind education: Retinitis Pigmentosa in Usher syndrome.* (videotape). Terre Haute: Blumberg Center for Interdisciplinary Studies in Education, Indiana State University. This 20-minute videotape presents the impact Retinitis Pigmentosa has on vision. A description of the progressive nature of the syndrome is presented. Actual pictures of a retina affected by retinitis pigmentosa are displayed. Additionally, behaviors that may indicate early signs of retinitis pigmentosa are identified including night blindness and tunnel vision.

Kansas Deaf-Blind Loan Library
Kansas Instructional Resource Center
Kansas School for the Blind, 1100 State Ave.
Kansas City, Kansas 66102
1-800-572-5463, ext. 5

Davenport S.L.H. (1996). *Important topics in deafblind education: Balance.* (videotape). Terre Haute: Blumberg Center for Interdisciplinary Studies in Education, Indiana State University. The 20-minute videotape illustrates anatomical functions that control balance including vision, muscle structure, hearing, the cerebellum, and joints. It takes the viewer through a journey of child motor development. This video also includes clips of children with various stages of CHARGE Association showing their chronological ages and their histories of motor and balance development

Kansas Deaf-Blind Loan Library
Kansas Instructional Resource Center
Kansas School for the Blind, 1100 State Ave.
Kansas City, Kansas 66102
1-800-572-5463, ext. 5

Davenport, S.L.H. (1996). *Important topics in deafblind education: Genetics and causes of birth defects* (videotape). Terre Haute: Blumberg Center for Interdisciplinary Studies in Education, Indiana State University. This 16-minute videotape presents conditions that can impact the genetic system during embryonic development including multifactorial elements, infections, narcotics and alcohol, diet, chromosomal, and the environment. Descriptions of steps a geneticist conducts to determine the location and type of syndrome are presented. Some infections related to deafblindness are described, such as Toxoplasmosis, Rubella, Cytomegalovirus, Herpes, and Syphilis (TORCH).

Kansas Deaf-Blind Loan Library
Kansas Instructional Resource Center
Kansas School for the Blind, 1100 State Ave.
Kansas City, Kansas 66102
1-800-572-5463, ext. 5

Edwards, L., Goehl, K.S., Gordon, L. & Houghton, J. (1992). **Profiles: Individuals with deaf-blindness**. Terre Haute: Indiana Deaf-Blind Services Project, Indiana State University.

This manual provides descriptions of common syndromes related to deaf-blindness. Each syndrome listed contains information on incidence, impact of the vision and hearing loss, physical descriptions, and personal testimonies of individuals who have the condition. This is a very powerful and informative manual that brings in the human component as well as the medical one.

Director of the Indiana Deaf-Blind Services Project
Blumberg Center for Interdisciplinary Studies in Education
SOE 502

Indiana State University
Terre Haute, IN 47809
(817) 237-2830

Enos, J. & Jordan, B. (1996) **A guide for students who are deaf-blind considering college**. Sands Point, NY: Helen Keller National Center-Technical Assistance Center. This guide was developed for students who are pursuing education beyond high school and for people assisting these students achieve their goals. The document helps students assess their own learning styles so they can better advocate for their needs. In addition, it helps them assess the strengths and weaknesses of potential post-secondary settings.

Helen Keller National Center
111 Middle Neck Road
Sands Point, NY 11050
(516) 944-8900 x307

Gallaudet College. (1978). **A screening program for Usher's syndrome**. Washington, DC: Author.

This manual presents a vision screening model for Usher syndrome that educationally assesses the person's visual field only. Other data gathering tools (e.g., family history, behavioral characteristics, and student interviews) are listed as necessary to complete screening activities. Although most screening models consist of additional types of evaluations related to sight, many of the suggestions and ideas in this manual are pertinent to screening sites.

Public Service Programs, Deaf-Blind Program
Gallaudet University
800 Florida Ave. NE
Washington, DC 20002
(202) 651-5051

Hennessey, J.C. (1991). **The inheritance of RP and allied retinal degenerative diseases**. Baltimore: National Retinitis Pigmentosa Foundation, Inc.

This booklet presents the genetic pattern of retinitis pigmentosa and the patterns of inheritance. Autosomal dominant, autosomal recessive, and sex-linked transmissions are described. Retinitis pigmentosa can occur in any one of the above listed patterns. A glossary of terms makes this document easy to read and understand. National Retinitis Pigmentosa Found., Inc.

d/b/a/ RP Foundation Fighting Blindness
1201 Mt. Royal Avenue, 4th Floor
Baltimore, MD 21217
Toll free: (800) 683-5555

Indiana Deaf-Blind Services Project. (1997, Draft). **What is Usher syndrome: How to recognize the combination of hearing less and retinitis pigmentosa**. Terre Haute: Blumberg Center for Interdisciplinary Studies, Indiana State University.

This manual describes conditions related to Usher syndrome using a question and answer format. It provides information related to behaviors often related to individuals with Usher syndrome as vision loss progresses. It discusses the importance of early detection, with descriptions of screening tests and ophthalmologic testing. Resources for learning more about Usher syndrome are located at the end of the manual.

Karen Goehl
Blumberg Center for Interdisciplinary Studies in Education
SOE 502
Indiana State University
Terre Haute, IN 47809
(812) 237-2830

Kansas Usher Screening Committee. (1996). **A collection of resources on Usher syndrome**. Topeka: Author.

This packet contains information on Usher syndrome from various state and national organizations. Additionally, there are two resource lists of specialists in the greater Wichita and Kansas City areas focusing on ophthalmological evaluations, genetic counseling, support services, and social-emotional counseling.

Director of the Kansas Deaf-Blind Project
Kansas State School for the Blind
1100 State Avenue
Kansas City, KS 66102-4411
Phone: 913-305-3014

Beth A. Jordan, Regional Representative CRC
Helen Keller National Center, Great Plains Regional Office
450 E. Park Street
Olathe, KS 66061
913-677-4562 voice
913-227-4282 videophone
Beth.Jordan@hknc.org

Kansas State School for the Deaf
Department of Nursing
450 East Park
Olathe, KS 66061
(913) 791-0573

Kansas State School for the Blind
Outreach Department
1100 State Ave.
Kansas City, KS 66102
(913) 281-3308 (Voice)

McGann, R. (1995). ***Tactile interpreters for students who are deaf and blind.*** (videotape). Gibsonia, PA: Distance Learning Center.

This two-hour videotape presents various strategies to use when communicating with someone who is deaf-blind including those individuals with Usher syndrome. The presenter demonstrates how to approach a person who is deaf-blind, assist a person who is deaf-blind, and end a conversation with a person who is deaf-blind. Tactile sign language is introduced, including components of this communication system. This video is accompanied by a booklet that highlights important aspects presented in the tape.

Kansas Deaf-Blind Loan Library
Kansas Instructional Resource Center
Kansas School for the Blind, 1100 State Ave.
Kansas City, Kansas 66102
1-800-572-5463, ext. 5

Minter, I., & Cioffi, J. (1997). ***Usher syndrome in the school setting.*** Sands Point, NY: Helen Keller National Center-Technical Assistance Center.

This resource presents issues around the identification, intervention, and supports needed for students with Usher syndrome. The authors stress that early intervention and contact with adults who have Usher syndrome are necessary if students are to feel good about themselves and have a sense of their futures. Strategies that assist in developing good self-esteem and essential life skills consist of participating in problem solving, sound decision-making, and introducing role models. Introduction to these strategies should occur early in the student's education as they can enhance successful life experiences as the disability progresses. This manual lists next steps, national and state resources, information and support regarding the education and rehabilitation of students with Usher syndrome.

Helen Keller National Center
111 Middle Neck Road
Sands Point, New York 11050
Voice (516) 944-8900 x 307

Sims, B., Raistrick, K., McGann, R., Schmidlin, R., Iurlano, A. & Baumgarner, J. (1996, March). ***When hearing loss and Retinitis Pigmentosa happen together: Meeting educational needs.*** (videotape). PennTech Teleconference Presentation, Gibsonia, PA: Education Service Center.

This three-hour video describes the symptoms for each type of Usher syndrome. Individuals with Usher syndrome talk about their experiences growing up with the condition. Accommodations and modifications needed when there is a decrease in the visual field are presented. National resources are described. A paper highlighting the issues discussed in the videotape accompanies the tape

Kansas Deaf-Blind Loan Library
Kansas Instructional Resource Center
Kansas School for the Blind, 1100 State Ave.
Kansas City, Kansas 66102
1-800-572-5463, ext. 5

Rothstein, R. (1998). ***The light at the end of The tunnel: Practical strategies for living with Usher syndrome.*** Sands Point, NY: Helen Keller National Center.

This book is written for individuals with Usher syndrome, but will also be helpful for relatives, friends, teachers, counselors, employers, co-workers, and service providers. The first section of the text explains Usher syndrome and its effect on individuals, including vision, hearing, balance, and genetics. The rest of the text looks at how to copy with Usher, including psycho-social and practical methodologies.

Much of the information has been gathered from people who have Usher syndrome. The publication also has a resource list of agencies, organizations, and information sources.

Helen Keller National Center
111 Middle Neck Road
Sands Point, NY 11050
Voice (516) 944-8900

Smith T., (1992). ***Deaf-blind communication and community: Overview and introduction.*** (videotape). Burtonsville, MD: Sign Media Inc.

This one-hour, open-captioned videotape discusses a number of topics relevant to communication for individuals who are deaf-blind with an emphasis in communication with individuals who have Usher syndrome. It contains clips of various individuals participating, traveling, and communicating within their community environments.

Kansas Deaf-Blind Loan Library
Kansas Deaf-Blind Loan Library
Kansas Instructional Resource Center
Kansas School for the Blind, 1100 State Ave.
Kansas City, Kansas 66102
1-800-572-5463, ext. 5

Smith, T. (1992). ***Deaf-blind communication and community getting involved: A conversation.*** (videotape). Burtonsville, MD: Sign Media Inc.

This 90-minute videotape presents live conversations with individuals who have Usher syndrome. They discuss how they learned they had Usher syndrome, changes they had to make in their lives once their vision deteriorated, and career opportunities. These individuals discuss the necessary supports and supplementary aids that assist individuals with Usher syndrome. Interpreters also share some of their experiences in working with individuals who have Usher syndrome.

Kansas Deaf-Blind Loan Library
Kansas Instructional Resource Center
Kansas School for the Blind, 1100 State Ave.
Kansas City, Kansas 66102
1-800-572-5463, ext. 5

NATIONAL RESOURCE AGENCIES

American Association of the Deaf-Blind (AADB)
814 Thayer Avenue, Suite 302
Silver Springs, MD 20910-4500
E-mail: aadb@erols.com

Boys Town National Research Hospital
Genetics Department
555 N. 30th Street
Omaha, NE 68131
(800) 835-1468

Dr. Sandra Davenport
Sensory Genetics/Neuro-development
5801 Southwood Drive
Bloomington, MN 55437-1739
(612) 831-5522

Fighting Blindness News
The Foundation for Fighting Blindness
National Retinitis Pigmentosa Foundation, Inc.
Executive Plaza 1, Suite 800
11350 McCormick Road
Hunt Valley, MD 21031-1014
Toll Free (800) 638-5555
Webpage: <http://www.blindness.org>

Helen Keller National Center (HKNC)
111 Middle Neck Road
Sands Point, NY 11050
(516) 944-8900
Webpage: <http://www.helenkeller.org>

Hereditary Deafness Newsletter of America
The National Research Register for Hereditary
Hearing Loss
Boys Town National Research Hospital
555 N. 30th Street
Omaha, NE 68131
(402) 498-6631

National Center on Deaf-Blindness
<https://nationaldb.org/>
Western Oregon University
345 N. Monmouth Ave.
Monmouth, OR 97361
E-mail: info@nationaldb.org

National Information for the Center on Deafness
(NICD)
Gallaudet University
800 Florida Avenue NE
Washington, DC 20002
(202) 651-5051

Retinitis Pigmentosa Foundation
1401 Mount Royal Avenue, 4th Floor
Baltimore, MD 21217-4245
(410) 225-9400

Usher Around the World
c/o Vision Screening Project
5801 Southwood Drive
Bloomington, MN 55437
(612) 831-5522

STATE & REGIONAL RESOURCES

Beth A. Jordan, Regional Representative CRC
Helen Keller National Center, Great Plains Regional Office
450 E. Park Street
Olathe, KS 66061
913-677-4562
913-227-4282 videophone
Beth.Jordan@hknc.org

Sensory Losses
<http://www.ksde.org/>
Kansas State Department of Education
Landon State Office Building
900 SW Jackson St, Suite 620
Topeka, KS 66612
Toll Free in Kansas (800) 203-9462
E-mail: jhoughton@ksde.org

Kansas Deaf-Blind Project
<http://kansasdeafblind.kssdb.org/>
Kansas State School for the Blind
1100 State Avenue
Kansas City, KS 66102-441
913-305-3014

White Canes & More
Vision Rehabilitation Center
530 N. Lorraine, Suite 100
Wichita, KS 67214
(316) 681-0870

COUNSELING

Ron Lybarger, Therapist
6142 Kenwood Avenue
Kansas City, MO 64110-3348
(816) 822-3679

Tag Goodspeed, Therapist

High Plains Mental Health Center
208 E. 7th St.
Hays, KS 67601
(785) 628-2871
John Kingsley, Therapist

ComCare of Sedgwick County
1919 N. Amidon St., Suite 130
Wichita, KS 67203
(316) 832-0318

OPHTHALMOLOGICAL EVALUATIONS

Dr. Trudi Grin, Ophthalmologist
10550 Quivira Road
Overland Park, KS 66215
(913) 888-1888

Dr. Gerhard Cibis, Pediatric Ophthalmologist
4620 JC Nichols Parkway
Kansas City, MO 64112
(816) 444-2943

University of Kansas Medical Center
Department of Ophthalmology
3901 Rainbow Boulevard
Kansas City, KS 66160
(913) 588-6600

Children's Mercy Hospital
Department of Ophthalmology
2401 Gillham Road
Kansas City, MO 64108
(816) 234-3000

Dr. Linda Buettner
Vision Rehabilitation Center
530 N. Lorraine, Suite 100
Wichita, KS 67214
(316) 682-4646

Dr. Leslie Nesmith, Ophthalmologist
530 N. Lorraine
Wichita, KS 67214
(316) 683-5611

Dr. Michael Varenhorst, Ophthalmologist,
Retina Specialist
530 N. Lorraine
Wichita, KS 67214
(316) 682-4646

Dr. David Waldie, OD,
Director of Optometry
Vision Rehabilitation Center
530 N. Lorraine, Suite 100
Wichita, KS 67214
(316) 682-4646

Dr. Paul Wieshaar
Ophthalmologist, Retina Specialist
530 N. Lorraine, Suite 100
Wichita, KS 67214
(316) 682-4646

GENETIC COUNSELING

Dr. David Harris, Geneticist
Children's Mercy Hospital
2401 Gillham Road
Kansas City, MO 641208
(316) 688-2080

Dr. Sechin Cho, Geneticist
Wesley Medical Center
550 N. Hillside
Wichita, KS 67214
(316) 688-2080

EQUIPMENT RESOURCES

Come Adaptation Test
Precision Vision
745 N. Howard
Villa Park, IL 60181
(630) 833-1454

What's Tunnel Vision? (simulators)
RP Research Foundation
366 Adelaide Street West, Suite 704
Toronto, Ontario, Canada M5V 1R9
(416) 598-4951

Appendix A: Supplemental Materials

What Is Retinitis Pigmentosa?

Retinitis Pigmentosa (RP) is an inherited condition. It affects the sensory cells (rods and cones) and the blood vessels of the retina. It may cause pigmentary clumping and a loss of electrical responses in the area of the eye. Usually the peripheral retina is affected first. This area of the retina contains the largest majority 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).

The cones are affected next. 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. Another condition that may be present which will affect central vision (Usher Type I) are cataracts. These may develop in the lens of the eye and may be present by the time a person reaches 20-40 years of age. Once cataracts have developed, a person may have difficulty seeing bright light, fine detail, and printed words. Everything may appear cloudy.

Characteristics of Usher Syndrome and Sensorineural Hearing Loss

DIFFERENT TYPES OF SENSORINEURAL HEARING LOSS		
TYPE	DECIBLE RANGE	WHAT CAN A PERSON HEAR
Normal	0-15 dB	Everything: all environmental sounds and speech sounds
Mild to Moderate	15-30 dB	Watch ticking, whispers & waterfall: sounds of f, th, s, z
Moderate	30-50 dB	Normal conversational speech but not the full spectrum of sounds: j, m, d, b, n, ng, e, i, a, o, r, p, h, g, ch, sh, k
Moderate to severe	50-70 dB	Air conditioner; loud voices; difficulty in group situations
Severe	70-90 dB	Piano, food blender, loud vowel sounds, lawn mower, telephone, dog barking
Profound	90-110 dB	Chair saw, semi truck, helicopter, train , airplane, very loud horn

CHARACTERISTICS ASSOCIATED WITH THE THREE TYPES OF USHER SYNDROME			
	TYPE I	TYPE II	TYPE III
HEARING LOSS	Congenital severe to profound bilateral sensorineural hearing loss; corner audiogram	Congenital moderate to profound bilateral sensorineural hearing loss; sloping audiogram	Normal hearing to moderate bilateral sensorineural hearing loss at birth; rapid progression of hearing loss for 5-10 years; sloping audiogram with ski bump
VISION LOSS	Early onset of blind spots; early onset of night blindness; peripheral vision loss; usually results in total blindness during adult years.	Blind spots or ring scotoma by teenage years; peripheral vision loss; usually does not progress to total vision loss	Blind spots or ring scotoma by teenage years; peripheral vision loss; progression varies but usually legally blind by young adult years
OTHER VISUALLY-RELATED CONDITIONS	Early development of cataracts	Varied central acuity loss	Progression varies but nighttime vision loss precedes daytime vision loss
BALANCE	Balance problems; walks at 18-24 months; does not get dizzy	Balance normal	May have some balance problems
GENETICS	1A long arm of 14; 1B long arm of 14 (most common); short arm of 11 (Acadian descent)	2A long arm; 2B long unknown	3A long arm of 3 (Finnish descent)

What Is an Electroretinogram (ERG) Evaluation?

An ERG is an evaluation that measures the electricity discharged by nerve impulses in the retina. This test is conducted by a board certified ophthalmologist. It can predict RP in children beginning at six to seven years old and for some children it can be predicted at younger ages (reportedly as young as three years old).

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 which will record responses of the retina. The person places his or her head in a globe called a Ganzfeld 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 one used in the skin electrode method. As in the skin electrode method, the responses made by the retina are recorded. The doctor will interpret the responses that indicate onset of RP.

Usher Screening Inservice Agenda

(names of presenters/coordinator of agenda)

(date)

(location---at prospective screening site)

15 min	Welcome & Introduction of Screening Team Members
30 min	What Is Usher Syndrome?
10 min	What Are We Doing in Kansas?

Screening for Usher Syndrome

20 min	Run Through a Typical Screening Day
10 min	Role Assignments
	Match Screeners to Stations
	How to Work with an Interpreter
20 min	Determine Station Locations
25 min	Role-Play Station
25 min	Cone Adaptation Station
25 min	Balance Station
25 min	Field Screening Station
5 min	Screening Expectations
5 min	Wrap-Up

How to Explain Usher Syndrome to Kids

Step 1: Briefly describe What happens with Usher Syndrome

“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 sign 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. There will be very little light in room.

“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 touch your legs and your eyes will be open. You will lift your arms up to the side like “you wanted to fly.” The adult will stand behind you and lightly touch the sides of your body (truck) 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 touch your legs and your eyes will be open. You will lift your arms up to the side like “you wanted 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 Field Test

“During the Field Test, you will sit directly across from an adult. You will be asked to sit 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 might 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 worried about how you did, tell the adult. Someone will be glad to help you.”

Appendix B: A Case Study

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 ASL and print for receptive and expressive communication. He possibly has some difficulties with vision, as noted by his teachers and parents. Sam was referred to his school nurse for educational Usher Screening.

In preparation for the station screening, the nurse sent the family history information to Sam's parents, behavior checklists 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 as follows:

Paper Screening: Family History

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 had worn glasses since the age of seven. Additionally, she marked several behaviors that indicated Sam was having difficulty seeing at night (e.g., difficulty seeing at night, 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 a geneticist.

Paper Screening: Student Interview

Sam reported that his eyes take longer than a minute to adjust to a darkened room. He stated that he sometimes trips over thing, 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: Behavior Checklist

Both Sam's mother and his social science teacher completed behavior checklists. There was a consistent pattern in night blindness, visual fields, balance, and other areas. There were inconsistencies reported for glare, contrast, and acuity.

Sam's audiogram revealed a corner audiogram consistent with audiograms that would indicate Usher syndrome I. He has a 90 dB loss that drops off dramatically in the lower frequencies.

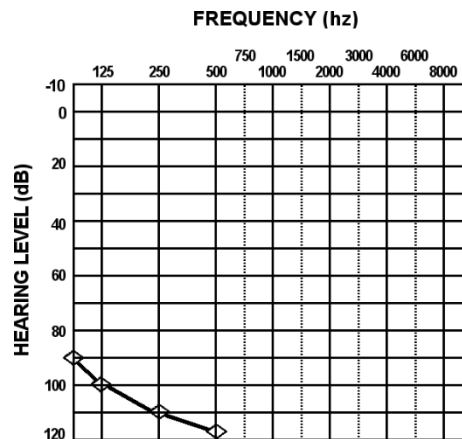
Cone Adaptation Station Screening

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

Sam failed 75% of the Balance Test. He only passed the section with His eyes open and both feet together.

Sam's visual fields were tested using both eyes. Both horizontal and vertical fields indicated a possible loss in the periphery.

Since Sam did not pass any of the station screening, his paper screening indicated visual field losses, balance difficulties, night blindness, and his family history indicated the presence of Usher syndrome, the school nurse referred Sam for an ERG and extensive ophthalmological evaluation with a physician with experience in visual field losses related to syndromes. The test was paid for by Sam's parents' insurance. Sam's mother currently is waiting for the test results.

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FAMILY QUESTIONNAIRE # 3			
OTHER HEALTHY TOBACCO USE			
Did the mother have pregnancy problems?		Yes	No
7 yrs. please check answers			
Problems during delivery?			
		Yes	No
Placenta: Birth weight: 7 lbs 7 oz Birth length: 20" Head size: 34 cm Circumference: 32 cm			
Did the mother have problems? (check one)			
1 per year please write number only		1	2
Did the baby have any problems like:			
1 per year please check one		1	2
	Birth problems		Breathing problems
	Malnutrition		Other: <u> </u>
How would you describe the baby's behavior?			
1 per year please write number in the hospital or home		1	2
	ALWAYS ON EMBURY		
0-100 = 0	100 = 100		
0-100 = 1	100 = 100		
0-100 = 1	100 = 100		
How many times did you use any tobacco products?			
1 per year please check one		1	2
	Yes		No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?		Yes	No
How many times have you visited the mother?			
1 per year please write number only		1	2

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KANSAS USHER SCREENING PROJECT

STUDENT QUESTIONNAIRE

Date of Birth: 10/24/89
Today's Date: 10/24/89

Discuss Sam Evans

How to Interview: Please take a minute to complete the questionnaire with your student or the student and the interviewer. This is to be completed for students from age 8 (or beginning in the first grade) through high school.

Discuss with David Ebyrd now every time he is the first to notice subtle change associated with grade to notice in all other children.

When you will make a shift from middle to teens, how long does it take you to get to a certain age, or middle, or teens?

When you come much from a bright, every day, it is hard to me things and people?

Yes

Does age make a difference?

Sometimes

Do you know your own age?

Yes, I wear short pants.

Do you also get people when passing them into a cup or glass?

Yes

Can you see the size of the day or night?

No

Is there a way to you from the size of the day, or you find when you are looking at them?

Yes

It is hard to find subtle things when you drop them on the floor?

Sometimes

Do you know your age, class, or things?

Sometimes

Do you have trouble seeing a circle?

Yes

I won't find one.

Do you have trouble seeing a glass of water or a glass of milk?

Sometimes

10/24/89

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Appendix C: Blank Forms

Usher Screening Forms (Summary Sheet)

Family Questionnaire

Student Questionnaire

Behavior Questionnaire

Cone Adaptation Test

Field Screening Test

Balance Test

Usher Screening Data Summary

Note: the user of this manual is granted permission to copy these forms.

KANSAS USHER SCREENING PROJECT

Student's Name: _____ Birth Date: _____

USHER SCREENING FORMS

Dates

1. Family Questionnaire

Completed _____

audiogram attached (if applicable) _____

2. Student Questionnaire _____

3. Behavior Observations

teacher _____

parent _____

dorm teacher _____

4. Cone Adaptation _____

5. Field Screening _____

6. Balance Screening _____

7. Additional Comments _____

KANSAS USHER SCREENING PROJECT

Usher Screening Project For Children Who Are Deaf or Hard of Hearing

FAMILY QUESTIONNAIRE

Name of your child	Birth Date	Today's Date
Address		
Phone #		
Name of person filling out this form:	Relation to student:	

QUESTIONS ABOUT CHILD

HEARING

How old was your child when he/she became deaf?	_____	Years
Why did he/she become deaf?		
Has he/she ever worn hearing aids?	Yes	No
If yes, did the hearing aids help?	Yes	No
Was your child's hearing ever better than it is right now?	Yes	No

VISION

Has your child ever worn glasses?	Yes	No
If yes, how old was your child when he/she got glasses?	_____	Years
Does he/she have problems seeing objects far away, but sees up close (nearsighted)?	Yes	No
Does your child have problems seeing up close, but sees well far away (farsighted)?	Yes	No
Has your child ever had eye drops put into the eyes to dilate them?	Yes	No
If yes, were there any problems with the drops? Please describe:		
Does he/she have difficulty seeing at night or in the dark?	Yes	No
Is he/she afraid of the dark or of shadows?	Yes	No
Does he/she have difficulty seeing the stars at night?	Yes	No
Does he/she have difficulty seeing sign language or gestures in dim light? (like at dusk)	Yes	No
Does he/she complain that bright lights hurt or bother him/her?	Yes	No
Does he/she need to wear sunglasses in order to see in bright sunlight?	Yes	No
When entering a new place or going from bright light to dim light or vice versa, does he/she ever stop suddenly, stand, and look around?	Yes	No
Does he/she ever confuse colors?		
If yes, is the color problem with red & green? ____ Yellow & blue? ____ Dark colors like navy, black, or brown? ____		

BALANCE

At what age did he/she sit? ____mo. crawl? ____mo. walk alone? ____mo.		
Did he/she have difficulty crawling or an abnormal crawl?	Yes	No
If yes, please describe how he/she crawled or moved about.		
Did he/she walk with the feet apart longer than most kids? If yes, until what age? ____mo.	Yes	No
When learning to walk, most children sit down hard on their seats and catch themselves with their hands if they start to fall. Did your child do anything differently? If yes, describe what happened.	Yes	No
Did he/she wear out lots of shoes or trip because he/she "shuffled" instead of picking up the feet?	Yes	No
Do you think he/she is clumsier than other kids the same age? If yes, please describe.	Yes	No
When your child was 3-5 years old and you were going from the car to the house at night, did he/she: ____stand or wait until someone came to take his/her hand? OR ____walk to the house by him/herself?		
Does he/she have problems now walking on bumpy ground?	Yes	No
Does he/she bump into objects like tables and chairs?	Yes	No
Does he/she often knock things over at mealtimes?	Yes	No
Does he/she stumble on stairs and curbs?	Yes	No
When he/she walks on a curb or 4-inch wide board, does he/she fall off? Don't know	Yes	No
Does he/she know how to ride a bicycle (two wheeler)? At what age did he/she learn how to ride? ____yrs. How long did he/she need training wheels? ____	Yes	No
Does he/she know how to swim?	Yes	No
Does he/she have problems seeing stripes on the bottom of the pool? Don't know	Yes	No

OTHER HEALTH HISTORY

Did the mother have pregnancy problems? If yes, please describe.		Yes	No
Problems during delivery? If yes, please describe.		Yes	No
Newborn: Birth weight: ___ lbs. ___ oz. Birth length: ___" Head size: ___"			
Was the child born prematurely (too early)? If yes, how many weeks early?		Yes	No
Did the baby have any problems like: ___ Yellow jaundice ___ Breathing problems ___ Meningitis ___ Other			
How long did the baby stay in the hospital after birth?		___	Days
Has your child ever been in the hospital? If yes:		Yes	No
DATE/AGE	ILLNESS OR INJURY		
Does anything like soap, lotion, medicine, etc. make your child's skin red or swollen? If yes, what?		Yes	No
Has he/she ever been treated for seizures?		Yes	No
Has he/she ever been treated for fainting spells?		Yes	No
List any medications he/she regularly takes.			
Does he/she have any of the following? ___ White patch of hair ___ Kidney problems ___ White patches of skin ___ Goiter (large thyroid gland) or lump on neck ___ Arthritis ___ Diabetes ___ Bones breaking easily ___ Cleft lip (hare lip)/cleft palate (hole in roof of mouth) ___ Extra fingers or toes ___ Lip pits or dimples ___ Different colored eyes ___ Heart murmur or defect			

QUESTIONS ABOUT FAMILY

These questions are about your child's blood relatives, not relatives by marriage or adoption. The word "relation" in the tables below mean how that person is related to your child, not to you. When you answer these questions, please think about both the mother's and father's sides of the family, including your child's brothers, sisters, aunts, uncles, grandparents, great-grandparents, and cousins. Please print clearly.

Ethnic background (voluntary; 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 trouble?

RELATION	KIND OF EYE PROBLEM	AGE WHEN PROBLEM STARTED
_____	_____	_____
_____	_____	_____
_____	_____	_____
_____	_____	_____

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

RELATION	KIND OF HEARING PROBLEM	AGE WHEN PROBLEM STARTED
_____	_____	_____
_____	_____	_____
_____	_____	_____
_____	_____	_____

Does anyone in the family have neurological problems like seizures, dizziness, fainting spells, cerebral palsy, brain damage, or any other problem with the brain, spinal cord, or nerves?

RELATION	KIND OF NEUROLOGICAL PROBLEM	AGE WHEN PROBLEM STARTED
_____	_____	_____
_____	_____	_____
_____	_____	_____
_____	_____	_____

Who in the family does your child look like? _____

Are your child's mother and father related to each other? That is, are they cousins (even distant cousins)? Yes _____
 No _____

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

If yes, who was the geneticist and where (address)? _____

Is there anything else we should know about your child?

Reprinted with permission from: Davenport, S. (unknown). Usher screening project. Bloomfield, MN: author.

KANSAS USHER SCREENING PROJECT

Usher Screening Project For Children Who Are Deaf or Hard of Hearing

STUDENT QUESTIONNAIRE

Student: _____

Date of Birth: _____

Today's Date: _____

Note to Interviewer: Please take a minute to complete this questionnaire with your student or the student and the interpreter. This is to be completed for students from age 8 (or beginning in the third grade) through high school.

Students with Usher syndrome may often be the first to notice subtle changes associated with gradual vision loss. *Ask the student:*

- When you walk inside a dark movie theater or room, how long does it take you to adjust (e.g., a couple of seconds, one minute, or longer)?
- When you come inside from a bright, sunny day, is it hard to see things and people?
- Do you trip over things?
- Does sunlight hurt your eyes?
- Do you often spill liquids when pouring them into a cup or glass?
- Can you see the stars in the sky at night?
- If someone waves to you from the side, do you see them when you are not looking at them?
- Is it hard to find small objects when you drop them on the floor?
- Do you bump into people, chairs, or things?
- Do you have trouble riding a bicycle?
- Do you sometimes knock over a glass of water or other objects on the table at mealtimes?
-

Reprinted with permission from: North Dakota School for the Deaf. (1996) Usher syndrome procedure. Devils Lake, ND: Author

KANSAS USHER SCREENING PROJECT

Usher Screening Project For Children Who Are Deaf or Hard of Hearing

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 3 minutes to complete.

Name of child: _____

Birth date: _____ **Current date:** _____

Completed by: _____

If you have noticed any of the following behaviors, put an "X" in front of the statement. 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 speaker
- _____ may express a desire to enter a room before it is darkened (e.g., movie theater)
- _____ avoids conversations in 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 Field

- _____ stumbles on stairs and curbs
- _____ bumps into people, tables, and chairs
- _____ may bump or tip over objects placed to the side at mealtimes
- _____ startles easily
- _____ seems to hold objects in unusual positions when looking at them
- _____ turns head while reading across a page
- _____ uses fingers to mark place while reading
- _____ can't find 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 lights 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 difficulty reading light copies or ditto copies
- _____ can't see stars at night
- _____ often spills when pouring liquids
- _____ eyes become watery and red when reading

Acuity

- _____ holds book close to eyes or bends to read
- _____ sits near blackboard

Balance

- _____ late learning to walk (past 15 months)
- _____ is considered clumsy
- _____ loses balance easily in the dark
- _____ can't ride a bicycle or required a long 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 at particular times
- _____ may fail to participate fully in group activities associated with new situation in the dark
- _____ frequently hesitates at the top or bottom of the stairs
- _____ avoids walking or running in unfamiliar areas, especially in bright sunlight or dark area
- _____ constantly appears to be visually scanning a group
- _____ Does your child wear glasses or contacts?

If yes, _____ for reading only? _____ for distance only? _____ all the time?

Adapted from: Illinois Usher Screening Project. (unknown). What you need to know about Usher syndrome. Springfield, IL. Author

Adapted from: Arizona Usher Screening Project (1995). Vision questionnaire. Tucson: Arizona School for the Deaf and Blind.

KANSAS USHER SCREENING PROJECT

Usher Screening Project For Children Who Are Deaf or Hard of Hearing

CONE ADAPTATION

Student's Name: _____ **Age:** _____

Date: _____

Procedure:

Scatter the squares from the Cone Adaptation Test in a jumbles heap on a dark table or carpet. The lights in the room should be dimmed to imitate twilight or dusk. The lighting needs to be even in the room. To see 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. Ask the student to pick up the white squares. Note how long the student takes to complete the task. Then ask the student to sort the red squares from the blue squares. Document the start time, stop time, and total time it took to complete the task. Document the number of squares correctly sorted. Conduct this test two times for each student.

Results:

Trial #1

Trial #2

_____ Time started

_____ Time started

_____ Timed stopped

_____ Timed stopped

_____ Total time

_____ Total time

_____ Number correct
(white) (blue) (red)

_____ Number correct
(white) (blue) (red)

Adapted from: North Dakota School for the Deaf (1996). Dark adaptation test. Devils Lake, ND: Author

KANSAS USHER SCREENING PROJECT

Usher Screening Project For Children Who Are Deaf or Hard of Hearing

FIELD SCREENING

Student's Name: _____ **Age:** _____

Date: _____

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

Wiggling Fingers Test

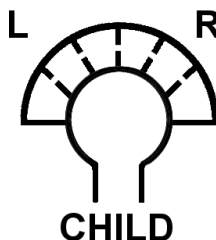
This test should be conducted with both of the student's eyes open. You and the student should be seated facing each other at approximately one arm's length. Tell the student to look at your nose and tell you when he or she sees your fingers. Extend both of your arms to the side of your body. Bend both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

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 at which point the student can see the fingers. Continue bringing your hands in closer toward your body to detect for ring scotomas. Conduct the test for both horizontal and vertical planes.

Results: 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.

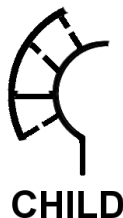
Horizontal



_____ both eyes

Results: 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.

Vertical



_____ both eyes

Adapted from Davenport, S. (1996). *Vision screening project*. Bloomington, MN. Author.

Adapted from: Alaska Center for Blind and Deaf Adults & Alaska Services for Children and Youth with Sensory Impairments. (1994).

KANSAS USHER SCREENING PROJECT

Usher Screening Project For Children Who Are Deaf or Hard of Hearing

BALANCE

Student's Name: _____ **Age:** _____

Date: _____

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 or her balance. Conduct the same procedure again, but this time the student's eyes need to be open. Document the student's reaction.

Balance maintained

Balance not maintained

eyes closed with feet together

eyes open with feet together

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 or her balance. Conduct the same procedure again, but this time the student's eyes need to be open. Document the student's reaction.

Balance maintained

Balance not maintained

eyes closed with one foot in front of the other

eyes open with one foot in front of the other

Adapted from: North Dakota School for the Deaf (1996). Balance. Devils Lake, ND: Author

KANSAS USHER SCREENING PROJECT

Usher Screening Project For Children Who Are Deaf or Hard of Hearing

USHER SCREENING DATA SUMMARY

Student	Birth date	Name of district or school
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Person completing data summary form	Date completed
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COMPLETED? WHY NOT?

Family Questionnaire	YES	NO
Student Questionnaire	YES	NO
Behavior Questionnaire form:	YES	NO
Parents	YES	NO
Teacher	YES	NO
Dorm teacher	YES	NO

Behaviors

COMPLETED?

Night blindness	YES	NO
Visual field	YES	NO
Glare	YES	NO
Contrast	YES	NO
Acuity	YES	NO
Balance	YES	NO
Other	YES	NO

COMPLETED?

APPEARS NORMAL?

Cone Adaptation	YES	NO	YES	NO
Field Screening	YES	NO	YES	NO
Balance Screening	YES	NO	YES	NO

RECOMMEND SENDING?

TO WHOM?

Referral letter	YES	NO
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COMPLETED?

TO WHOM?

Followup	YES	NO
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Comments: