

Usher Syndrome

Introduction

Usher Syndrome is an autosomal recessive genetic disorder characterized by congenital hearing loss and gradually developing retinitis pigmentosa leading to the loss of vision. Approximately 25,000 people in the United States have some form of Usher Syndrome. Most of these individuals have either Type I (10,000) or Type II (15,000).

Type I Usher Syndrome is characterized by profound congenital deafness, vision problems starting in early childhood, and severe balance problems.

An individual born with a moderate to severe hearing loss and normal balance characterizes Type II Usher Syndrome. Night blindness for Type II begins at childhood, but many individuals might not be aware of it until late adolescence/early adulthood.

Historically, most students with Usher Syndrome Type I attended residential schools and colleges for the deaf, while individuals with Type II attended regular public schools and universities. With the mainstreaming of deaf and hard-of-hearing students brought about by Public Law 94-142, students with both Type I and II Usher Syndrome can be found in all types of educational settings. It is critical for support service personnel and counselors in college/university environments who work with students with Usher Syndrome—regardless of type—to have a good understanding of special issues that arise for these students and to be aware of strategies that will benefit this population. It also is critical that professionals have a good understanding of Usher Syndrome and what it means to be deaf/hard of hearing and to be losing one's vision.

College-age students with Usher Syndrome have many of the same developmental issues as other adolescents (age 18 to 24 years). However, students in this population have additional issues that are not encountered by students who have normal hearing and vision or even their deaf and hard-of-hearing peers who have normal vision. The purpose of this tipsheet is to identify particular issues of concern for

college-age students who have Usher Syndrome, and list some strategies and tips for the reader. The reader will also find a list of resources for both professionals and students.

Counseling Issues for Students with Usher Syndrome

Educational Issues:

Students will want to know:

- What major/career is best for them?
- Can they continue in this career for several years? How do they know how long they will be able to function in their chosen career if the medical profession cannot predict the progressive deterioration of their vision?
- How to educate teachers and others about classroom needs without bringing special attention to themselves?
- What can they do to help themselves find their way around the campus at night?
- What strategies do they need to learn to adapt to changes in vision and how it affects the educational process?
- What adaptive equipment is necessary or helpful, e.g. Closed Circuit TV for computers and reading books, special Corning CPF glasses that will reduce glare, and use of yellow or orange transparencies in the classroom?

Personal/Social and Life Issues:

- How will the student interact with peers when communication in groups and in dark places is difficult, due to diminishing dark adaptation and shrinking visual field?
- How will the student adapt to changes in vision and hearing?
- What special concerns will the student have related to dating and relationships?
- Can anyone love a person with Usher Syndrome? What is the impact of Usher Syndrome on marriage?
- Can the student have children; will he or she need genetic counseling?
- Will the student be able to raise children properly?

- How can the student talk with his/her parents if the family never discusses Usher Syndrome?
- What are some of the positive and negative experiences of people who have Usher Syndrome?
- Where can role models be found?
- How does the Americans With Disabilities Act apply to the student's needs in the educational/work environment?
- What organizations, services, and professionals can help the student move from being a deaf/hard-of-hearing person with vision limitations to becoming legally blind?
- How can the student deal with issues related to anger, guilt, shame, depression, and fear of dependency?
- What if the student does not want to discuss or even acknowledge his/her Usher Syndrome?

Strategies and Tips for Working with College-Age Students Who Have Usher Syndrome

The first rule of thumb is never to assume what the student does or does not know about Usher Syndrome. Assess his or her knowledge about the medical aspects as well as what he or she knows about services available to people who have Usher Syndrome.

For many students, it often is easier to be more open about their educational needs than their personal-social needs, so this is a good place to start. As a support service professional or counselor, your first priority should be to do a *needs assessment interview* with the student. Find out what he/she needs from you. Develop a list of questions related to educational issues such as:

- Do you have trouble finding your way around in new places? Can you hear or see the fire alarm in your dorm room? Can you see in dimly lit places?
- Do you have trouble reading the blackboard? Is it easier for you to read whiteboards with black markers?
- Do you have a problem with glare with overhead projectors or in the classroom?
- Do you have difficulty reading regular printed materials? Does it help to have a larger font size such as 18 point or 24 point? (Show the student examples of different font sizes.)
- Are you able to follow your interpreter in the classroom? Is his or her use of space outside of your vision range?

- Do you have any concerns about your chosen career and your future?

Strategies for Educational Issues

1. The student might need mobility and orientation training on campus and in the surrounding community. Locate these services in your community by contacting the Commission for the Blind and Visually Handicapped (CBVH). If there are no services available in your community, then work with the student and his or her family to see if these services are available in the family's hometown. Another option is to discuss with the student the possibility of attending a summer program at the Helen Keller National Center for young adults. You should be working with the student's Vocational Rehabilitation Counselor or the Commission for the Blind and Visually Handicapped to secure these services and payment of services.
2. Some students can read blackboards easier and others prefer the whiteboard with black markers. Red, green, and blue markers are more difficult to read. If your school does not have these options then use of a notetaker would be critical.
3. Use of an interpreter is very important for those students who use sign language. If the student is attending a college where faculty and students all use sign language, then use of a copy interpreter (someone who sits within the visual range of the student and repeats what has been signed or spoken) could be very beneficial.
4. If the student is in a mainstream setting, utilize an interpreter and notetaker. If the student is oral, then a notetaker or C-Print technology would be of great benefit.
5. Use 18- or 24-point font on overhead projectors.
6. Use yellow transparencies to reduce glare.
7. Give the student hard copies of information from transparencies.
8. Good lighting in the classroom is essential.
9. Use good contrast with media, e.g. black on white or black on yellow or dark blue with white fonts.
10. Investigate the possibility of the student getting a CCTV (closed circuit television) for reading books and helping with enlarging prints on the computer. If the student needs this type of equipment for success in the classroom, he/she

will need it for success in life as well. Work with the student's sponsoring agency for funds.

11. Do research and educate yourself about careers of people who are deaf/blind to assist students with concerns or confusion about career choice. It is hard to predict how each person's vision might change as they age. Some will become totally blind by the age of 40 and others might keep a good part of their central vision until they are in their 70s. It is not the counselor's role to tell students what career to choose. The counselor should help the student understand that many people change career paths three or four times in their lifetime. Encourage the student to do research about careers. Also encourage the student to learn as much as possible about Usher Syndrome and how it might influence the future. Currently one can find people with Usher Syndrome working as:

Researcher, professor/author, chef/owner of restaurant, lawyer, certified public accountant, information technology specialist, computer programmer, public health manager, counselor/ professor, minister, librarian, veterinarian, health planner, clerk, medical transcriptionist, mental health counselor, high school teacher

Strategies for Personal/Social/Life Issues

1. Often a student will not be emotionally ready to deal with having Usher Syndrome. If the student is functioning well academically and personally, leave the denial alone. Denial is an important aspect of how people protect themselves mentally until they are ready emotionally to deal with having Usher Syndrome. Of course this often makes it hard to work with students who are not functioning well academically or emotionally because of their Usher Syndrome. In this case, it is important to gently try to help the student understand how his/her changing vision or lack of vision is having a serious impact on how he/she functions in the classroom or with peers. If a student does not want to talk about these issues, it is best to leave him/her alone. This might lead to classroom failure, but a failure could be the catalyst leading to change.
2. Sometimes acceptance is not possible if the visual problems are not yet apparent.
3. If the student is having problems communicating in group settings or dark places, encourage the student to be more open about having Usher

Syndrome and talking about his or her communication needs with close friends.

4. Encourage the student to meet other people who have Usher Syndrome (role models), especially adults who have learned how to adapt and make modifications in their life to function well and independently. This can be done through the Internet and also by contacting organizations that involve people with Usher Syndrome. (See resources list.)
5. Find out about organizations, e.g. Helen Keller National Center, and websites that will help you as a professional but also help your student. (See resources list.)
6. If possible, try to find a person who has Usher Syndrome who can function as a role model and mentor for your student. We all know how important it is for deaf and hard-of-hearing students to have role models and peer groups. The same is true for students who have Usher Syndrome.
7. If a student is depressed, angry, and afraid, suggest counseling services. If you feel you are not qualified to work with clients on those issues, seek out a qualified therapist to whom you can refer the student.
8. Consider encouraging the student to attend a special summer program at Helen Keller National Center for adolescents and young adults. This program will give the student an opportunity to be with peers who have Usher Syndrome and meet older adults who have Usher Syndrome. It also will give students an overview of career possibilities, orientation and mobility, independent living skills, and computer and communication adaptive technology, and will help them to be more independent and assertive about their needs.
9. Do not assume that all problems the student might have are the result of having Usher Syndrome.
10. Offer hope and be honest. If the student asks, "Will I go blind?" the answer is, "I do not know." No one knows what will happen for any particular person.
11. Be open, accessible, and approachable to the student.

Resources

DBTeens is a private email Internet forum for teens and young adults to share information, ideas, opinions, and other issues for people who have Usher

Syndrome or are deaf/blind. To subscribe, contact owner Ralph Klumph at klumphr@wou.edu. Send the message "subscribe DBTeen."

Usher Syndrome List

An Internet forum for people with Usher, their parents, friends, spouses, and service providers. To join this list via Yahoo! Groups, go to <http://groups.yahoo.com/group/Ushers/join>. To subscribe, click on [Join](#).

Organizations that provide information or services

American Association of the Deaf/Blind

814 Thayer Avenue
Silver Spring, MD 20910
301-588-6545 (T)
301-588-5705 (V)
aadb@erols.com

Center for the Study and Treatment of Usher Syndrome

Boys Town National Research Register for Hereditary Hearing Disorders
(National Institute on Deafness and Other Communication Disorders)
555 North 30th Street
Omaha, NE 68131-9909
800-835-1468 (V/T)
402-498-6331 (F)
genetics@boystown.org

DB-LINK

The National Information Clearinghouse on Children Who Are Deaf-Blind
Teaching Research Division
Western Oregon University
345 N. Monmouth Avenue
Monmouth, OR 97361
800-438-9376 (V)
800-854-7013 (T)
503-838-8150 (F)
www.tr.wou.edu/dblink
dblink@tr.wou.edu

The Foundation Fighting Blindness

Executive Plaza I, Suite 800
11350 McCormick Road
Hunt Valley, MD 21031-1014
888-394-3937 (V)
410-785-1414 (V)
800-683-5551 (T)
410-785-9687 (T)
410-771-9470 (F)
www.blindness.org

Helen Keller National Center for Deaf-Blind Youths and Adults (HKNC)

111 Middle Neck Road
Sands Point, NY 11050-1299
516-944-8900 (V)
516-944-8637 (T)
516-944-7302 (F)
www.helenkeller.org
hknctrng@aol.com

These are Web site links that will connect you to other resources and various Web sites that have information about Usher Syndrome as well as deafness/blindness in general.

www.tr.wou.edu/dblink/links.htm

www.tr.wou.edu/dblink/source.htm

For more information, contact:

Northeast Technical Assistance Center

Rochester Institute of Technology

National Technical Institute for the Deaf
52 Lomb Memorial Drive
Rochester, NY 14623-5604

585-475-6433 (V/TTY)
585-475-7660 (Fax)
Email: netac@rit.edu
<http://netac.rit.edu>



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