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Usher Syndrome: Systematic Review of the Literature, Case Studies, and Recommendations for Future Research

Kathleen Ceaser, Stacy O'Connell, and Theodore Scott Smith

Usher syndrome represents a comparatively rare genetic disorder, resulting in both hearing loss and visual impairment. Rehabilitation planning may facilitate not only improved activities of daily living, but also economic self sufficiency, and employment. To make possible employment for persons with Usher syndrome, the rehabilitation professional must understand basic etiology surrounding the syndrome, and also prospective worksite modifications needed to promote vocational independence. This article offers a general summary of the history and symptoms of the syndrome, assisting the rehabilitation professional with rudimentary knowledge. Moreover, a listing of the current literature is offered, alongside specific references, country of origin, and publication type, aiding and, perhaps, prompting additional research on this topic. Two case studies are further offered, leading the rehabilitation professional to apply presented information on this topic. Supplementary recommendations are presented to direct both short-term and long-term research on Usher syndrome.

Usher syndrome (US) represents an autosomal recessive genetic condition that affects hearing, vision, and in some cases, balance (Annala, 1983; Boughman, Vernon, & Shaver, 1983; Fishman, Kumar, Joseph, Torok & Andersonj, 1983). Among these patients, hearing is most often the initial presenting symptom (Astuto et al., 2002; Brown, 1987; Dreyer et al., 2001; Emmorey, Korpics, & Petronio, 2009). As vision is affected by US, similarly, equilibrium may be affected. More specifically, the symptoms of US are reflective of not only hearing loss, but also Retinitis Pigmentosa (RP) (Fakin, Zupan, Glavac, & Hawlina, 2012; Grøndahl, 1987; Sadeghi, Cohn, Kimberling, Halvarsson, & Moller, 2013). The vision loss is termed Retinitis Pigmentosa which results from the progressive degeneration of the retina; typically, with first presentation as night-blindness, with supplementary loss of peripheral vision, and eventual demarcation to what is commonly termed as "tunnel vision." Interestingly, while the history of US equates to the early 1900's (Usher, 1914; Vernon & Duncan, 1990; Vernon, 1996), research over the past two decades on US has been quite sparse. Anecdotally, based on experience of the first two authors, rehabilitation professionals are often unadvised, unaware, and badly informed about basic knowledge pertaining to US. As such, this article seeks to discuss the symptoms and subtypes, general etiology, assessment, and assistive technology applicable to US. Furthermore, two case studies are presented, offering the rehabilitation professional an opportunity to apply the information and facts presented in the present paper.

History

Charles Usher, a Scottish ophthalmologist, initially deemed a syndrome characterized with poor vision and hearing as "usher syndrome" in the mid 1910s. He had observed sixty-nine cases in which there appeared to be a genetic link between parents and offspring relating to the actual identification and presentation of visual and hearing impairments in children. While Charles Usher was initially credited with being the originator of the syndrome, Usher syndrome type disorders were identified as early as the mid 1850s. A pioneer of modern ophthalmology, Albrecht von Grafe, reported a genetic link between two brothers who had relatively the same symptoms, specifically Retinitis Pigmentosa (von Gräfe, 1858). Again, in the mid part of the 1800s, Richard Liebreich began to examine congenital patterns among individuals with deafness, specifically those individuals with Retinitis

Pigmentosa (Liebreich, 1861). During this period of time, the main medical perspective was to identify a genetic link as opposed to actual treatment and education. It was not until the mid 1900s that education, training, and eventual employment were considered for individuals with Usher syndrome.

Differential Diagnoses

As will be reflected throughout the present paper, Usher syndrome is incurable at present. It is recommended that efforts be taken to diagnose children early, particularly before they develop night-blindness. It has been suggested that as many as 10% of deaf children with congenital linkages may have Usher syndrome. Differential diagnoses may have bad results and incorrect treatment protocols. Particular differential diagnoses may include Alport syndrome, Cockayne syndrome, Flynn-Aird syndrome, and Hurler syndrome.

Client Education & Worksite Accommodations

From an educational standpoint, there are several tips or recommendations for students or workers with Usher syndrome. When developing short-term and long-term vocational planning, the rehabilitation professional must be aware of worksite accommodations, materials, techniques to improve communication, and also self advocacy. Pertaining to worksite accommodations, one should be advised that lighting should be adequate to offer a full view of a room or worksite, without glare. Perhaps, utilization of full spectrum lighting would facilitate visual adaptations for individuals with minimal or reduced visual capabilities. Furthermore, employers should provide group instruction in a non-cluttered area to not only avoid unneeded movement, but also to avoid the likelihood for an individual to stumble and cause injury. Again, reflecting the worksite environment, windows should be behind the students or employees to avoid glare. If chalkboards or whiteboards are used in a work setting, these media should be cleared of unnecessary or unneeded marks in order for the individual with Usher syndrome to differentiate important material from miscellaneous or worthless information. Furthermore, regular printing as opposed to cursive lettering may be used to identify narrative content.

Correct selection of learning media would be vital towards necessary accommodations within a worksite. Perhaps the rehabilitation professional, such as the vocational counselor, may conduct a learning media assessment to identify not only the applicable learning media within a work environment, but also strategies to accommodate the disability. If written materials are provided, the rehabilitation professional may consider that the person with Usher syndrome may need individual copies of charts or graphs, as these individuals may require additional time to both examine and comprehend the selected materials. If employment or educational testing is utilized, the rehabilitation professional may consider the need for test adaptations such as alternative forms and also assurance that test booklets or test materials are in the necessary font size.

Oftentimes, individuals with Usher syndrome may utilize sign language or sign interpreters (Tactile sign may be used if loss of vision is severe). For individuals with limited, but sufficient vision, printed signage within a worksite must be not only of sufficient size to be read, but also as concise as possible in order to avoid extraneous data. Utilization of some type of interpretation services may be necessary for business meetings or continuing education courses that would allow not only access but also interaction with selected materials to ensure comprehension.

Due to the nature of the disability, time allowances may be needed for individuals with Usher syndrome within multiple work-based situations. The typical framework for educational testing is that individuals receive "time and a half" to complete comparable amounts of work as their peers. The rehabilitation counselor may consider speaking with an employer about allowing additional time or reducing work requirements to allow for successful completion of the work task. Special efforts must be taken to ensure that an individual is not excluded from a particular work environment based upon the nature of the disability.

Most importantly, the rehabilitation professional must consider self advocacy as a necessary component of vocational planning. The individual with Usher syndrome must develop strategies to not only discuss necessary adaptations and accommodations needed for themselves for an immediate work task, but also projected for long-term needs. Psychological issues may represent concurrent symptoms, further requiring mental health consultations (Hess-Rover, Crichton, & Holland, 1999; Rijavec & Grubic, 2009; Waldeck, Wyszynski, & Medalia, 2001; Wu, & Chiu, 2006).

Symptoms and Subtypes

While the diagnosis of Usher focuses on the loss of hearing and vision, it is diagnostically more complicated than equating the syndrome to simply a hearing and visual disorder. It represents a complicated classification scheme and has psychosocial implications as per separate classifications (Miner, 1997; Cohen, Bitner-Glindzics, & Luxon, 2007; Cote, Dube, St-Onge, & Beauregard, 2013; Mets, Young, Pass, & Lasky, 2000; Vernon, 1974; Vernon, 1978). There are three types of Usher syndrome: Type I, Type II, and

Type III. The different types vary pertaining to both characteristics of the person with a disability, and severity of the disability itself (Avraham, 2013; Cremers et al., 2007; Ebermann et al., 2010; Garcia-Garcia et al., 2011; Green, Yang, Grati, Kachar, & Bhat, 2013; Otterstede, Spandau, Blankenagel, Kimberling, & Reisser, 2001). Type III represents the least common type of US in the United States; Types I and II represent the majority of US distribution, equating to almost 95 percent of all reported occurrences. The following paragraphs represent discussions pertaining to type of US, alongside specific diagnosis criteria.

Individuals diagnosed with Type I are born profoundly deaf (Cremers et al., 2007; Dahl et al., 1993; Dreyer et al., 2001; Kimberling et al., 2010; Miner, 1995). That is, these individuals have a hearing loss of 91 decibels or higher, resulting in speech that is not only inaudible, but also the reality that hearing aids typically offer little benefit or improvement in hearing abilities. Furthermore, balance problems tend to manifest themselves, particularly during the first few years of life, specifically as the child is learning to sit up and crawl. The noted balance problems manifested during the infant and toddler years most often carry along towards the adult years. Individuals with Type I tend to experience the onset of RP toward the end of their first decade, or approximately at 10-12 years of age (Cohen, Bitner-Glindzicz, Luxon, 2007). Type I has been typified as the most severe pertaining to global hearing loss. Considering the presence of hearing loss and vestibular (balance) issues at birth, coupled with the progressive vision loss over the next 1-2 decades, Type I requires continual adaptations for not only the person with the disability, but also the family unit.

It has been suggested that Usher Type II is associated with fewer auditory and visual severity features than Type I (Miner, 1997). Most often, it is characterized as having a congenital origination, resulting in moderate to severe hearing loss, and concurrently offering a diagnosis of RP, typically manifested in late adolescence through a person's mid-twenties. It has been proposed that Type II represents the most prevalent form of Usher, accounting for more than half of all cases (Cohen, Bitner-Glindzicz, Luxon, 2007). While people with Type I are typically born profoundly deaf, those with Type II have a moderately severe hearing impairment. Moderately severe hearing loss is defined as a 56 – 70 dB loss; in this range, speech is inaudible without amplification. However, with hearing aids, speech may be heard, but not necessarily understood or interpretable by the person with US. Typically, with Type II, vestibular function is not affected, resulting in lack of impairment in balance and gait progression.

Usher Type III is a more recently discovered classification, with classification originating within the last 20 years (Miner, 1997). Individuals with Type III have a variable onset of progressive hearing loss and vision loss. That is, a person may not definitively develop hearing and vision losses during the first decade of life, but may develop representative losses later in life, perhaps during their teenage years, up to one's 20's. Typically, a person with Type III may develop hearing loss by their teens and may require hearing aids by their 20's (NIDCD Fact Sheet: Usher Syndrome. Publication No. 98-4291). In addition, night-blindness, which occasionally begins during puberty and usually by mid-adulthood, may also accompany a Type III diagnosis. Most importantly, in Types I and II, hearing loss precedes RP; however, with Type III, vision loss may occur prior to hearing loss.

Diagnosis and Testing

As noted previously, US represents an autosomal, recessive genetic condition (Oshima et al., 2008; Petit, 2001; van Wijk et al., 2004; Xu et al., 2011; Zhou et al., 2012). It is passed down from carriers of the mutated gene. If both parents carry the mutated gene, they will have a one-in-four chance of having a child with Usher. If parents are aware that they have the gene, they can act accordingly when the child is born by planning for both educational and activities of daily living accommodations. Across children with US, parents most often will notice their child's hearing loss within the first 2-4 years of life. Upon recognizing the hearing deficits in their children, parents will bring the child in for testing. An audiological evaluation will typically be performed, perhaps prompting the child to be diagnosed with deafness or hearing loss, depending on both the type of US, and the severity of sensational loss. Again, depending on the type, Usher Syndrome is usually confirmed once RP manifests itself.

There are many tests that can be performed to diagnose RP including a visual field test which will indicate the field of vision. As mentioned earlier, night blindness and a narrowing of the field of vision represent some of the symptoms that prompt parents or individuals themselves to seek testing. Other testing for RP may include an electroretinography (ERG) which tests to see if the retina is working properly and an electrooculogram (EOG) which measures electrical impulses from the back of the eye to the front.

Treatment and Assistive Devices

There is currently no cure for Usher Syndrome (Astuto et al., 2002; Dahl et al., 1993; Dammeyer, 2012; Domanico et al., 2012). As such, the best treatment options are intended to assist the individual with their residual hearing and vision, coupled with improving activities of daily living and vocation independence (Duncan, Prickett, Finkelstein, Vernon, & Hollingsworth, 1988; Emmorey, Korpics, & Petronio, 2009; Vernon, 1969; Williams, 2008). Of course,

assistive devices, such as hearing aids and amplification devices can assist with improvement of hearing capabilities, if there is indeed residual hearing. Different hearing aid options may be available for these individuals. Individuals may choose to wear hearing aids in their ear canals, behind their ears, or surgically embedded in their skulls (Cochlear Implants). Beyond the use of aids to increase the function of the ear, there are other devices that can assist the individual in certain settings. For example, the person may be a student and an FM Device may offer the most benefit. This is a device that is worn around the neck (or sometimes connected via Bluetooth) by the hard of hearing person. The teacher or lecturer will wear a microphone around his/her neck so that what is said will be transmitted to the coil or loop around the student's neck. The loop will then transmit the sound into the hearing aid. A very common way to assist those with hearing loss is the use of interpreters. If the individual has learned sign language and relies on that for communication, having an interpreter present to provide access into the communication is priceless. There are a few different forms of sign language that are utilized.

When the vision of the individual has been drastically reduced by Retinitis Pigmentosa, he or she will need to learn many new skills for living an independent life. These skills and the training involved are not in the scope of this article, but will be addressed in future articles. Some forms of treatment/assistance for people who are deafblind include glasses, bioptics, high-power magnifiers, and software/devices that convert text to speech. Once the individual has lost all usable vision, they must rely on their cane or a guide dog to remain mobile. Cue cards can be made in advance that can be used to assist the individual with Usher syndrome to communicate with others. For example, cue cards may be used to ask for assistance in crossing the street or for directions. Adaptive aids may also be used to assist in communication. Companies such as Humanware develop and sell products that make communication possible for individuals with Usher syndrome. The Deafblind Communicator (DBC) offers assistance in three different areas of communication including face-to-face, TTY, and SMS texting. The DBC has made communication and independence possible for many individuals with Usher syndrome and other forms of deafblindness.

Mobility impairment is associated with the diagnosis of Usher Syndrome. It is crucial that the individual receive adequate training in Orientation and Mobility (O&M) to insure the best possible mobility outcomes. Blindfold training is often used to teach the individual with Usher syndrome who may have some residual vision skills that they will need when they lose more vision. It may be proposed that crossing signs have been shown to be effective in the safe crossing of streets and intersections for deafblind individuals. The crossing signs request that pedestrians or passing drivers assist the deafblind individual to cross the street, they may read "I am deafblind, please tap me if you can help me."

Systematic Review of the Literature

For the present review, the first and third authors identified 35 articles, utilizing nationally, available databases, including PsychLit, PsychInfo, and Ebsco. In summary, the authors utilized 15 journal abstracts, and 20 full text journals in their current review. The literature is broken down as follows; 15 peer reviewed journal abstracts, 14 full-text peer reviewed journal articles related to Usher Syndrome and genetics, 2 full-text peer reviewed journal articles related to psychosocial aspects of Usher Syndrome, 2 full-text peer reviewed journal articles related to Usher Syndrome and psychosis, and 2 full-text peer reviewed journal articles related to Usher Syndrome in children (early diagnosis and behavioral disorders). The authors utilized the following research and organized it by topic and specialty. See Table 1 for a breakdown of the literature.

The majority of the research that has been published on Usher syndrome has been done by medical doctors with a focus on genetics. Aside from organizations for the blind and deafblind such as the Helen Keller National Center, Association for Education and Rehabilitation of the Blind and Visually Impaired, Seattle Lighthouse for the Blind, National Institute on Deafness and Other Communication Disorders, and Affiliated Blind of Louisiana Training Center, there is a lack of published research available for rehabilitation professionals related to rehabilitation and employment of persons with Usher syndrome.

Case Studies

The following case studies are being presented as an example of the psychosocial implications that individuals diagnosed with Usher syndrome may experience.

Walter, 15, Usher Syndrome Type 2

Walter was born with moderate to severe hearing loss. He attended a state school for the deaf and communicates via American Sign Language (ASL). At the age of 15 Walter began to experience night blindness which prevented him from being able to drive at night or get around in the dark without bumping into things. At the age of 18 Walter began college and majored in accounting with the goal of becoming a CPA. Shortly after starting college, Walter began noticing problems with his peripheral vision. He began noticing that he could not see other students who were sitting off to the sides in the stadium classrooms. Walter

also got into a car accident when he switched lanes and side swiped a car he stated was in his blind spot. His passenger informed him that she could easily see the other car in her peripheral view.

By this time Walter was having problems with receptive ASL as the signer had to be directly in front of him for him to be able to see the signs. The night blindness and increasing tunnel vision prompted Walter to see an Ophthalmologist who eventually (after several tests) diagnosed Walter with Usher syndrome type 2. After learning of his diagnosis, Walter became clinically depressed and isolated himself in his apartment. He contemplated leaving college and moving back home with his parents rationalizing that he would never be able to live independently and work to support himself. As his vision loss (retinitis pigmentosa) increased even more Walter was no longer able to adequately communicate with his deaf and hard of hearing peers via ASL.

Walter sought counseling from a counselor who was experienced in working with individuals with disabilities including blind, deaf, and deaf-blind individuals. Eventually Walter began using tactile sign language in order to communicate and utilizing accommodations including assistive technology in order to finish college and eventually be employed as a CPA. Walter used calculators and computers with a Braille display. He makes use a of device called a deaf blind communicator (DBC) which converts text to Braille and Braille to text on a mobile device to communicate with his employer, co-workers, and customers.

Amelia, 22, Usher Syndrome Type 1

Amelia was the second child to her parents, Tessa and Jack. Their first child, Jack Jr., was born hearing and with good vision. Amelia was diagnosed with profound sensorineural hearing loss at age four and was not fitted with hearing aids because the doctor felt they would not benefit her. The doctor recommended they send Amelia to a residential school for the Deaf so she could learn sign language and be a part of the Deaf Culture. Tessa and Jack did not want to send their daughter far away to school, so instead, they moved to Baton Rouge to live near the School for the Deaf. They saw Amelia regularly during the week and every weekend.

Amelia went to school and learned American Sign Language (ASL); her parents and Jack, Jr. learned it also so they could communicate comfortably with their family member. Amelia grew up in a supportive family at a school with other Deaf children. She was not treated differently than other kids and grew up with a strong sense of self and also pride in her community. At this time, she developed an interest in computers and working with numbers. She began to focus on this career goal. When Amelia turned 16, her mother realized that she had stopped asking about a driver's license. When she questioned Amelia, she learned that she was having trouble seeing at night and she was afraid to get behind the wheel of a car. Tessa brought her in for an eye exam and after several tests and a few long weeks; Amelia was diagnosed with Usher Syndrome Type I. After interviewing Amelia, the doctor learned that she had started experiencing night-blindness about age 13 and by 15; her peripheral vision was closing in.

Amelia finished her schooling at the residential program, but began pulling away from her friends and the community she had built there. Her mother sent her to counseling. Through therapy, Amelia admitted that she was pulling away from friends because her Deaf peers wouldn't want to use tactile signing and they would ignore her. She had seen it happen with other people and she knew it would happen to her. Therapy did assist Amelia with her issues surrounding her diagnosis.

After graduation, Amelia was sent to an adaptive training center where the students learn to live independently with their vision loss, and in her case, her hearing loss as well. She learned Orientation and Mobility skills: how to get around with a cane and communication cards. She began to learn Braille and how to work with software and hardware that assist with her dual sensory loss. Amelia remained there for the duration of the 9 month program and once she completed it, she was contacted by a VR counselor to discuss her options. Amelia had always wanted to work on computers and after vocational assessments, it was determined that she would do well in data entry. Her computer, phone, and work space could be adapted to accommodate her dual sensory loss. Both Amelia and her counselor knew that her vision would deteriorate and that accommodations would be needed throughout her career. Amelia understood that Usher Syndrome was not an immediate loss to be dealt with and move on. Rather, it was a continuous movement of adapting to new realities. Because of her support systems that were in place, her education, training, and a VR counselor who understood her disability, Amelia was able to create a fulfilling life for herself.

Summary and Recommendations

Usher syndrome represents a significant challenge for the rehabilitation professional. The challenge relates to both understanding the syndrome itself and identifying proper assistive technology in vocational options. An initial step towards providing quality rehabilitation services is understanding the diagnosis.

One of the main areas that vocational rehabilitation counselors must become familiar is the utilization of assistive technology. Assistive technology may be utilized to not only assist with independent living, but

also within the workforce. Properly matching the assistive technology with the individual would be vital towards facilitating independent vocational independence. It would be recommended that a "cookie cutter" approach not be taken when assigning assistive technology, but rather the worker with Usher syndrome be consulted and become a vital part of the discussion process.

While a tremendous amount of research was generated at the beginning of the century and the middle part of the previous century, within the last two decades there has been a paucity of research pertaining to Usher syndrome. Reasons for the paucity of research may vary from lack of interest on behalf of the researchers themselves, funding, or a combination of these. However, while there has been a paucity of research, it is important to note that present records suggest that the number of individuals with Usher syndrome has not decreased. That is, the need to provide rehabilitation services continues, albeit the availability of empirically based research is not available. It is recommended that not only qualitative, but also quantitative research be generated over the next two to three decades to assist the rehabilitation professional in providing said services.

It is important that counselors be trained in assessment and recommending treatment regarding the specialized needs of clients with US (and other types of deafblindness). Counselors need to be knowledgeable about the different communication modalities for communication with deafblind individuals such as tactile sign language, print on palm (if sign language is not known), or other communication devices such as the deafblind communication (DBC). Individuals with Usher syndrome that work usually require specialized assistive technology that enables them to communicate with other as well as perform job duties. It is important that rehabilitation counselors be familiar with the various types of assistive technology that can be utilized by persons with Usher syndrome. It is important that deafblind consumer's be properly trained in using their assistive devices. Peer reviewed literature on the topics is needed to educate rehabilitation counselors who may work with consumers with Usher syndrome or other types of deafblindness. It should be noted that there is much more extended dialogue beyond the scope of the present article, but some of these modalities/devices are being offered to educate the reader.

The rehabilitation professional must take into account the prospective psychological factors affecting individuals with Usher syndrome. Psychological symptoms may include depression, or secondary psychological illnesses. As a result of the psychological overlay, the rehabilitation professional may consider the viability and need for a variety of professionals, including psychologists, psychiatrists, or a multitude of other mental health care providers. The rehabilitation professional may consider the provisional psychological services not only in the context of Usher syndrome itself, but also psychological needs that may or may not occur. When providing vocational services, the vocational counselor must consider the context of secondary disabilities. Most likely, the rehabilitation professional in a forensic setting will consider or review assistive technology and additional services whenever another illness or injury has occurred, such as within the context of a worker's compensation or motor vehicle accident. It may be difficult to separate work related components unless the components are disparate. That is, mobility associated with poor muscle condition and function may be easily separated from mobility impairment due to poor vision. It is important to note that mobility may be impaired with Usher syndrome without presence of a secondary disability, which makes such assessments even more complicated.

In addition to the physical features associated with Usher syndrome, the rehabilitation professional must consider the psychosocial issues. Psychosocial issues pertain to the need for an individual with Usher syndrome to not only understand themselves, but also work with others as a societal unit. The individual with Usher syndrome must work with other employees, their respective supervisor, and family members. Such impairments may include the inability to communicate through visual cues, poor mobility to access community or social events, and limited ability to hear. Furthermore, various assistive technologies may or may not be accepted within particular family structures according to family dynamics or cultural domains. As such, the vocational counselor must be aware of the psychosocial implications associated with this syndrome.

In summary, US represents a complicated syndrome, with a series of both physical and psychological overlay features. Depending on severity, individuals with US may, indeed, be employers. For the forensic rehabilitation professional, public rehabilitation professional, and the state rehabilitation professional knowledge about the syndrome is necessary to develop and modify rehabilitation plans. As always, general knowledge about the disorder must be accompanied by input by the client and patient, directly developing the rehabilitation plan. It is suggested that research in this area must be revitalized to accommodate the continued needs for these clients in the modern rehabilitation community.

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Table 1
Listing of Available Research on Usher Syndrome

Type of Publication	Publication Topic	Publication Specialty	Author's country of Origin/Date of Publication
Abstract (from book)	Behavioral & Psychological Treatment of Physical Illness	Psychology	U.S.A /1988
Abstract (from book)	Rehabilitation involving deaf-blind consumers	Rehabilitation	U.S.A /1990
Abstract (from Journal)	Clinical case study on the relationship between Usher and psychosis	Journal for the Study of Interpersonal Processes, Vol 64(3)	U.S.A /2001
Abstract (from Journal)	Clinical case study in Usher syndrome and psychiatric symptoms	Psychiatria Danubina, Vol 21(1)	Croatia /2009
Abstract (from Journal)	Empirical study of an intervention plan to help persons with Usher syndrome 2 adapt to deaf blindness	British Journal of Visual Impairment, Vol 31(2)	Canada /2013
Abstract (from Journal)	Empirical study on the use of visual feedback from evidence from signers with impaired vision	Journal of Deaf Studies and Deaf Education, Vol 14(1)	U.S.A /2009
Abstract (from Journal)	Differential diagnosis, Eye disorders, Genetic disorders, Hearing disorders, Mutations	Vision Research, Vol 75	Slovenia /2012
Abstract (from Journal)	Axons, Myelin Sheath, Proteins, Cytoskeleton, Animal Models, Mice	BMC Neuroscience, Vol 14	U.S.A/2013
Abstract (from Journal)	Hearing Disorders, Vestibular Apparatus, Drug Therapy, Mutations	The New England Journal of Medicine, Vol 369(18)	Israel/2013
Abstract (from Journal)	Expressivity of hearing loss in Usher type 2A	International Journal of Audiology, Vol 52(12)	Sweden/U.S.A/2013
Abstract (from Journal)	Psychological and Clinical Disorders	Journal of Chronic Diseases, Vol. 22(10)	U.S.A/1969
Abstract (from Journal)	Overview of Usher syndrome	The Volta Review, Vol. 76(2)	U.S.A/1974
Abstract (from Journal)	Psychosocial implications of Usher syndrome	American Annals of the Deaf, Vol 123(3)	U.S.A/1978
Abstract (from Journal)	Empirical study in Usher syndrome and communication techniques	Reading in Deafness, Mono 6	U.S.A/1983
Abstract (from Journal)	Demographics related to Louisiana students with Usher Syndrome	Journal of Visual Impairment & Blindness, Vol 81(3)	U.S.A/1987

Full-text Journal	Early diagnosis of Usher syndrome in children	Transactions of the American Ophthalmological Society. Vol. 98	U.S.A/2000
Full-text Journal	Mental and behavioral disorders in children with Usher syndrome	Psychiatry Dammeyer Behavioral Brain Functions	Denmark/2012
Full-text Journal	Psychotic illness in an individual with Usher syndrome	Journal of Intellectual Disability Research. Vol, 43	Germany, UK/1999
Full-text Journal	Usher syndrome and psychotic symptoms	Psychiatry and Clinical Neurosciences, 60(5)	Taiwan/2006
Full-text Journal	Usher syndrome: animal models, retinal function of Usher proteins, and prospects for gene therapy	Pharmacology and Neurosciences. NIH. Vision Res. Author manuscript	U.S.A/2008
Full-text Journal	Medical/Genetics related to Usher syndrome	Journal of Medical Genetics, 44	The Netherlands, U.S.A, Italy, Switzerland, UK, Belgium/2006
Full-text Journal	Medical/Genetics related to Usher syndrome	Molecular Vision	China/2011
Full-text Journal	Medical/Genetics related to Usher syndrome	Orphanet Journal of Rare Diseases	Spain/2011
Full-text Journal	Medical/Genetics related to Usher syndrome	Hum Mutat. (Human Mutation)	Spain, Japan, Sweden, U.S.A/2008
Full-text Journal	Medical/Genetics related to Usher syndrome	The American Journal of Human Genetics	The Netherlands/2004
Full-text Journal	Medical/Genetics related to Usher syndrome	Case Reports in Ophthalmological Medicine, Vol 2012	Italy/2012
Full-text Journal	Medical/Genetics related to Usher syndrome	The Journal of Clinical Investigation, Vol 120	Germany, U.S.A, Canada, France/2010
Full-text Journal	Medical/Genetics related to Usher syndrome	Molecular Vision	Canada, China, U.S.A/2012
Full-text Journal	Medical/Genetics related to Usher	American Journal of Human Genetics	U.S.A, The Netherlands, Spain, South Africa, France, Pakistan, Sweden/2002
Full-text Journal	Medical/Genetics related to Usher	American Journal of Human Genetics	Norway, Denmark, Sweden, U.S.A, Spain/2001
Full-text Journal	Medical/Genetics related to Usher	Journal of Medical Genetics	U.S.A, Sweden/1993
Full-text Journal	Medical/Genetics related to Usher	Genetics in Medicine	U.S.A/2010
Full-text Journal	Medical/Genetics related to Usher	International Journal of Audiology	U.K/2007

Full-text Journal	Psychosocial implications of Usher 1	Journal of Visual Impairment & Blindness. Vol 89	U.S.A/1995
Full-text Journal	Psychosocial implications of Usher 2	Journal of Visual Impairment & Blindness Vol.91	U.S.A/1997

The authors would like to recommend the following websites which can be utilized for further research on the topic of Usher syndrome:

Helen Keller National Center
http://www.hknc.org/WhoWeServeUsher.htm

Seattle Light House for the Blind
http://www.deafblindlh.org/

National Institute on Deafness and Other Communication Disorders
http://www.nidcd.nih.gov/health/hearing/usher.htm

Classification	**External Resources**
ICD-10	H35.5
OMIM	276900, 276901
DiseasesDB	13611
MeSH	D052245

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