

Hearing and Visual Impairments in People with Intellectual and Developmental Disabilities

Objectives

- What does it mean to be Blind, Deaf or DeafBlind
- How best to communicate with people with sensory impairments
- Strategies for assisting people with sensory impairments to have optimal participation in their health care



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- People who are deaf, blind or deafblind often have associated physical, intellectual, behavior, medical or motor problems
- Effective communication requires putting hearing and visual impairments in context with all the other problems
- Effective communication means learning the best means of communication with each individual, remember each individual is unique

Every Picture Tells a Story



- **Visual Impairments**

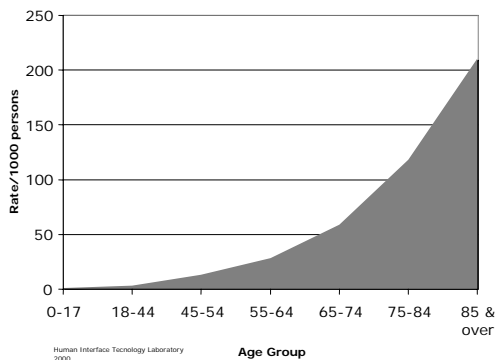
Gallup Poll 1989

- 42% of Americans polled said that blindness is “the worst thing that can happen”
- 71% of Americans age 45 years and older fear being blind more than being deaf
- 76% fear being blind more than having to use a wheelchair

Causes of Visual Impairments

- Structural: damage to one or more parts of the eye, examples cataracts, glaucoma, amblyopia, strabismus, retinal detachment
- Refractive Errors: inability of the eye to sharply focus the image on the back of the retina, examples myopia, hyperopia, astigmatism, anisometropia
- Cortical Visual Impairment: damage to the part of the brain that interprets visual information

Visual Impairments in the U.S.



Components of Vision

- Visual acuity: the ability to resolve detail, normal 20/20 (testing distance/letter size)
- Visual fields:
 - Reflects the health of the retina (central & peripheral areas of the retina)
 - Can have scotomas (blind spots) of any size & shape
 - Scotomas can be central or peripheral
 - Overall peripheral field defect can interfere with mobility
 - People with visual field loss need O&M and visual scanning training

WHO Classifications: The International Classification of Impairments, Disabilities and Handicaps (ICIDH)

- Visual impairment can be defined generally as any loss or abnormality of an anatomical structure or of a physiological or psychological function.
- Examples of visual impairments are: reduced visual acuity, visual field loss and loss of contrast.
- Visual impairments may be categorized as normal, near-normal, moderate, severe/profound, near-total or total vision loss depending on the degree of loss of visual acuity or visual field.

- Contrast:
 - When contrast sensitivity decreases it may feel like vision has worsened although measured acuity is the same
 - Testing gives indication of who will respond poorly to standard magnification levels or who will need higher levels of light needed for tasks
- Lighting and Glare:
 - Lighting that does not contribute to retinal imagery has an adverse affect on visual efficiency, comfort or resolution
 - Glare can be caused by the eye (cataracts) or externally

Legally Blind

- Legally blind means with the best correction visual acuity is 20/200 or worse in the better eye or if the remaining central visual field is restricted to 20° or less in the widest meridian of the better eye
- This definition dates back to 1935 when the Social Security act when its benefits to the blind was passed, the visual field part was added the following year

Vision Loss Consequences

- Loss of independence in ADL's
- Restrictions in mobility without training
- Limits access to education, recreation, employment, socialization
- Environmental safety concerns
- Sense of or actual isolation
- Depression

Common Causes of Visual Impairment Prenatal – Perinatal – Postnatal

- Genetic: Down Syndrome, CHARGE Syndrome, Usher Syndrome, Septo-Optic Dysplasia (Optic Nerve Hypoplasia), Retinitis Pigmentosa
- Prematurity: Retinopathy of Prematurity (commonly also associated with spastic diplegia)
- Hypoxia
- Infection: prenatal Rubella, CMV, Herpes, Meningitis
- Hydrocephalus
- Trauma

Information to Get From an Eye Exam

- Get a good explanation of the nature of the visual impairment and how it affects daily activities
- Practical size of the materials used for learning, reading, and doing ADL's
- Best lighting
- Explanations of the prescription for corrective lenses so that they are used for the appropriate activities
- Are there other devices that would be helpful

Common Causes of Visual Impairments Adulthood and Aging

- 80% of people with visual impairments are age 65 or older
- Cataracts: 50% of Americans age 65-74 and 70% over age 75 have cataracts
- Macular Degeneration
- Diabetic Retinopathy
- Glaucoma
- Get an eye exam if you notice new onset of stumbling, bumping into things, avoiding activities, loss of skills, anxiety in unfamiliar settings, difficulty on stairs or in poorly lit areas or night time

Cortical Visual Impairment

- Impaired or reduced vision due to neurological abnormalities
- There are specialized areas of the brain for distance vision, recognition of faces, objects, colors, contrast and movement, perceptions of directionality & depth, and hand-eye coordination
- Caused by: hypoxia, brain mal-development, head trauma or infection
- People with CVI are rarely totally blind and sometimes vision may improve although the cause does not resolve
- May co-exist with ocular visual loss, but usually normal optical exam

Common Visual Characteristics of CVI

- Vision may be variable or delayed
- Children may tend to use peripheral vision more effectively than central vision
- Some are photophobic, others are light-gazers
- Poor depth perception
- Close viewing is common
- Difficulty with complex visual images

• Hearing Impairments

Getting Information If You Are Blind

- Hearing-voice and noises
- Smells
- Touching objects
- Being touched by others
- Sensations-movement of air, vibrations
- Residual vision-light and dark, shadows, shapes or movement

Hearing Loss

- Normal hearing: -10-25 decibels
- Mild hearing loss: 26-40 decibels
- Moderate hearing loss: 41-55 decibel loss
 - Usually need hearing aids or other amplification to understand speech
- Moderately severe hearing loss: 56-70 decibels
- Severe hearing loss: 71-90 decibel loss
 - Requires special education with prolonged auditory training, speech and language and communication skills,
 - Benefits from hearing aids and other amplification methods
- Deaf - Profound bilateral hearing loss: >90 decibel loss
 - Relies totally on vision or other senses for communication, amplification typically not helpful

(normal conversation is 60 decibels)

Safety

- Avoid low hanging things such as plants, lights or signs
- Avoid clutter or excessive furniture
- Don't change furniture arrangements
- Good natural lighting, additional lighting will depend upon needs of the person
- Avoid things that cause glare such as glass or glossy posters
- Avoid sudden changes in light from bright to dark areas
- Use color contrasts, large letters and Braille
- Provide verbal information about the environment and obstacles

Types of Hearing Loss

- **Conductive hearing loss**
 - Affects the outer and middle ear
- **Sensorineural hearing loss**
 - Affects the inner ear
- **Mixed hearing loss**
 - Includes both conductive and sensorineural hearing loss

Common Causes of Hearing Loss Prenatal – Perinatal – Postnatal

- Genetic: Down Syndrome (50-75% have some degree of hearing loss), Usher Syndrome, CHARGE Syndrome, Laurence-Moon-Biedle Syndrome
- Hypoxia: often associated CP and seizure disorders
- Infections: Prenatal Rubella, CMV or Herpes, meningitis
- Recurrent Ear Infections
- Ototoxic Drugs: furosemide, aminoglycoside, some chemotherapy agents, aspirin causes tinnitus

Hearing Assistive Devices

Hearing Aids

- Amplify all sounds including your voice
- New technology can reduce much of the background noise
- Several varieties with constant improvements in technology
- Takes time to adapt and many people wear them only for specific situations
- Infants tend to adapt more easily than young children

Assistive Listening Devices, FM devices

- Shorten distance between listener & sound
- Brings sound directly to the ear
- Less background noise
- Good for theater, telephone, 1:1 conversation, radio/TV

Hearing Loss: Young to Old

Any age: noise, ear wax impaction

Aging:

- The 3rd most common chronic health problem in older Americans
- 40% of people age 65 have a hearing loss
- 80% of people age 85 and older have a hearing loss

Most commonly there is damage to the cochlea in the inner ear.

Cochlear Implants

- Electronic devices in which electrodes are surgically placed in the cochlea in order to stimulate the auditory nerve directly
- Main candidates are young children, adolescents and adults who developed some language prior to the development of the profound hearing loss (FDA approved minimal age requirement is 12 months, but there are exceptions for younger children)

Signs of Hearing Loss

- Muffled quality of speech and other sounds
- Difficulty understanding words, especially against background noise or in a crowd of people
- Asking others to speak more slowly, clearly and loudly
- Needing to turn up the volume of the television or radio
- Withdrawal from conversations
- Avoidance of some social settings

Additional Devices and Communication Techniques

- Alerting devices: Lights or vibrations
- TV closed captioning
- TTY: text telephone
- American Sign Language (ASL)
- Lip reading, fingerspelling
- Pictures, gestures, touch

Getting the Most Out of an Audiological Evaluation

Preparing for the evaluation:

- Ear check before the appointment for wax or infection
- Bring all equipment
- Bring a list of all medications, diagnoses, questions and concerns

• DeafBlindness

Information to Get Out of the Evaluation

- Review results for each ear
- What are the educational, conversational and safety considerations
- Would the person benefit from equipment
- Are there accommodations that need to be made in the setting
- Ideas and strategies to help the person get the most out of communication and learning

Deafblind Definitions

- The US Federal definition of deafblindness is “concomitant hearing and visual impairments, the combination of which creates such severe communication and other developmental and educational needs that they cannot be accommodated in special education in programs solely for children with deafness or children with blindness.”

FR Dept. of Education, 34 CFR Parts 300 & 303. Vol. 64, No.48.3/12/99.

Is a Hearing Aid Appropriate and Useful for Someone Who May Not Easily Tolerate One ?

- Benefits of hearing aids: assists with communication, allows the person to enjoy music or other sounds, makes the person more aware and safe in their environment
- Does not need to be worn all the time to have benefit
- Desensitization: start with an ear mold; a slow process, but possibly may result small but increasing amount of time the hearing aid is tolerated

Deafblind

- Deafblind-a combined loss of vision and hearing that significantly affects access to communication, learning, socialization, activities of daily living and mobility*
* (visionAustralia)
- When you think about deafblind you may think about a total absence of sight and hearing
- However, usually there is a varying degree of vision and hearing loss. The most important feature to remember is that it is the combination of loss that limits access to vision and auditory stimulation and natural opportunities to learn to communicate

Courtesy of NEC

Eligibility Chart and Classification Codes

	Degree of Vision Loss						Critical Vision Impairment (CVI)
	Normal 20/10 - 20/40	Near Normal 20/40 - 20/70	Low Vision 20/70 - 20/200	Legally Blind 20/200+	Legally Blind with Light Perception	Totally Blind or no Light Perception	
Degree of Hearing Loss							
Normal 0-25 dB							
Mild 26-40 dB		A			C		*
Moderate 41-55 dB			B		D		*
Moderate Severe 56-70 dB							*
Severe 71-90 dB					E		*
Profound 91 dB or greater							*

Facts About Deafblindness

- There are approximately 80,000 individuals ages birth to 65 in the U.S. with some form of deafblindness
- 10,000 are children
- 60% have physical impairments
- 68% have cognitive impairments
- 40% have complex health needs
- More than 90% of children with deafblindness have one or more additional disability or health problem
- The population with deafblindness has become more severely involved due to advances in medical technology that have improved survival rates for premature and seriously medically involved infants

- *For a deafblind person, the world literally shrinks in size and scope, and whatever knowledge is obtained must come through the secondary senses of taste, touch, and smell, and the exercise of personal curiosity and initiative. . . **

* N. Robbins in *Remarkable Conversations* (1999)

Deafblind Etiologies

- No known etiology-17%
- Hereditary syndromes-13%
- Prematurity-13%
- CHARGE syndrome-12%
- Pre-natal/Congenital-12%
- Pre-natal/Non-congenital-6% plus
- Microcephaly, Cytomegalovirus, asphyxia, hydrocephaly, Down syndrome, severe head injury, Usher syndrome, maternal drug use, Rubella syndrome, encephalitis
- The above conditions account for about 70% of the deafblind population*

*2006 National Deaf-Blind Child Count

Deafblind Demographics

- There are approximately 70 causes of deafblindness
- It can occur at birth; prematurity, childbirth complications, congenital syndromes (many rare), CHARGE syndrome*
- It can occur during childhood and adulthood due to meningitis, brain injury or inherited conditions*

* From National Consortium on Deaf-Blindness (2007)

- *Blindness takes an individual away from things, and deafness takes him away from people . . . Deafblindness creates unique problems of communication, mobility, and orientation, peculiarly its own.**

*N. Robbins in *Remarkable Conversations* (1999)

Getting Information If You Are Deafblind

- Tactile sign (ASL, signed English, etc.)
- Print (braille)
- Objects
- Tactile cues
- With residual hearing or vision-gestures, pictures, symbols, voice, social stories (signed and or pictures), large print, technology

As a Medical Professional for a Sensory Impaired Individual

- Ask questions of the individual if you can communicate with them
- Ask questions of care givers only if you can not communicate with the patient
- Read the profile if one is available, if not ask about communication skills and strategies
- Raising your voice with a person who has a sensory impairment is usually not helpful
- Slow down and think about how they are perceiving you

- *I can only talk with one person at a time, and only if that person is within my reach. If I am in a crowded room, and no one is within 3 feet of me, I might as well be in a closet. It is very easy for me to misunderstand, so communication is often very slow.**

* N. Robbins in *Remarkable Conversations* (1999)

Working with People Sensory Impairments in Health Care Settings: Hearing and/or Visual Impairments

- Identify yourself immediately
- Determine the person's preferred strategies for communication
- Don't rush, give the person a chance to process the information
- Eliminate any distracting noises
- Tell the person what you plan to do and how it will feel before each step
- Remember that the person has had many experiences in medical environments, some positive, but some have caused anxiety and stress
- Do not make assumptions about the assistance the person needs, ask, offer, don't just do

As an Advocate for a Sensory Impaired Individual:

Outline a profile for medical visits

- Take it with you to appointments
- The Profile should include:
 - Expressive communication skills and strategies
 - Receptive communication skills and strategies
 - Equipment use and non-use
- Speak up before the visit starts, don't wait until there is a problem

Working with People Sensory Impairments in Health Care Settings: Visual Impairments

- Describe the environment to the person before or as entering it, guide the person's hand to chairs, examining tables, sinks, toilets, toilet paper holders
- Allow the person to explore the room to get oriented, remember the blind person sees with his/her hands
- When walking with the person offer your elbow and walk in front of the person, describing doorways, stairs etc... just before you reach them (sighted guide)
- Never leave the person in free space
- Try not to move around too much so the person can stay oriented to you

- Described what is going on around the person, what you are doing away from the person
- Avoid descriptions that include shapes, patterns, colors, designs
- Announce when you or others enter or are about to leave the area
- Explain any written instructions (posted, handouts, signs)
- Keep the person informed and aware of the setting and activities using clear precise descriptions
- Do not pet or interact with guide dogs

Areas of Care Coordination for Children and Adolescents: Medical, Physical

- Growth and Development
- Nutrition
- Eating and Swallowing
- Dental care
- Hearing and vision
- Associated Medical Disorders including:
 - Neurological and Seizure Management
 - Renal
 - Endocrine
 - Immunological
 - Metabolic
- Neuromuscular - Motor Development – Cerebral Palsy
- Genetic counseling

Working with People Sensory Impairments in Health Care Settings: Hearing Impairments

- People who are deaf are legally entitled to the services of a qualified sign language interpreter
- Family members should be avoided as interpreters if there are concerns of confidentiality
- People with profound deafness pre-lingually (before acquiring speech) may have very different skills and culture than those who are late-deafened
- Always face the person and always talk to the person, even if there is an interpreter

Care Coordination: Psychosocial, Educational, Vocational

- Cognitive/intellectual status
- Behavioral, social and emotional concerns: ADD, ADHD, autistic like behaviors, oppositional behaviors, social and emotional skills training and assistance
- Orientation and mobility instruction
- Modified teaching strategies to accommodate for sensory impairments Opportunities for sports, recreation, social and leisure activities
- Opportunities for vocational assistance supports Need for OT/PT/SLP/specialized educational and vocational training/ADL training
- Speech and communication
- Need for adaptive equipment, assistive technology, aids

Care Coordination

- Many causes of hearing or vision loss are associated with other medical or physical disorders including cerebral palsy, seizure disorders, intellectual or developmental disabilities
- Genetic disorders may have associated physical, behavioral, nutritional, metabolic, immunological or endocrine disorders
- Hearing or vision loss may be a result of a medical condition such as stroke, diabetes, infection
- Often the diagnosis may have implications regarding management, progression, prognosis, rehab and related problems that need to be considered

The Sensory Profile

- Describe the sensory impairment
- Preferred mode of communication, both receptively and expressively
- Back up mode of communication if you do not know my primary mode
- What happens when I take off my glasses, hearing aids, cochlear, etc.
- Tips: special things that are helpful to me when I visit a medical professional

Example of a Sensory Profile for a Blind Individual

- I am a totally blind person with mild developmental disability. I understand simple spoken English, 4 or 5 words at a time. I can follow simple 2 step directions if you are specific.
- I prefer you to talk to me rather than my staff.
- If you ask a simple question I can respond. I have a reliable “yes” and “no” response.
- I prefer you to use speech both expressively and receptively.
- Please tell me what you are doing before you do it.

- Please say what you are doing before you do it. The interpreter will let me know. Give us a minute to communicate.
- If my interpreter needs to leave the room for privacy, please use written notes to communicate with me.
- It would be helpful if you could sign “Are you OK?”, “Ready” and “Finished”.
- When the interpreter re-enters the room tell me what the diagnosis is and the interpreter will sign to me. I may have questions that I will sign to the interpreter but she will voice for me to you. Tell me the answers.

- Please tell me before, how (hand or instrument) and where you will be touching me.
- It can be helpful if you ask me “Are you ready?”
- If you are looking for something, tell me what you are doing i.e., “I’m looking for a tongue blade in the drawer.”
- I have a very good sense of smell. If you are using liquids let me know because I will smell them.
- I need you to place my hand on the examination table and my chair when I need to move.

Example of a Sensory Profile for a Deafblind Individual

- I am a deafblind person with mild developmental disability. I can see some light and shadow and hear loud noises. I can not hear speech. I understand simple signed English, 4 or 5 words at a time from my staff who will sign into my hands. I can follow simple one step directions from my staff.
- If you ask a simple question and wait for my staff to sign to me, I can respond. I have a reliable “yes” and “no” response..
- Please tell my staff what you are doing before you do it. The staff will tell me. Give us time to communicate.

Example of a Sensory Profile for a Deaf Individual

- I am a totally deaf person with no cognitive limitations. I communicate through ASL with other deaf and hard of hearing individuals. When I go to a medical appointment I have an interpreter.
- I prefer you to look at and speak to me rather than the interpreter. I will get info from the interpreter but I want to see your face too.
- When you need to ask a question speak directly to me. I will answer in ASL and the interpreter will voice for me.

- Please tell my staff before, how (with your hand or an instrument) and where you will be touching me and the staff will tell me.
- It can be helpful if you ask me, through my staff, “Are you ready?”
- I have a very good sense of smell. If you are using fluids tell my staff so the staff can tell me.
- If my staff person is not next to me, I need you to place my hand on the examination table and my chair when I need to move. I move though space using sighted guide; this means I will place my hand above your elbow and you will guide me through space. I will be a little behind you. Watch out for doorways and furniture.

The Sensory Profile for _____

Date: _____

Person with the individual: _____

Relationship to the individual: _____

Hearing _____

Vision _____

Preferred Mode of Communication _____

Back Up Mode of Communication _____

If I must take off my glasses, _____


If I must take off my hearing aids, _____

If I must take off my cochlear _____

Tips to communicate effectively with me:

3. This treatment also can be paired with modeling e.g. watching someone else remaining relaxed in the situation (Love et al., 1990)
4. Present an item from the situation list starting with the least anxiety-provoking to the most anxiety-provoking while the person remains deeply relaxed

You are going to give Helen Keller, the world's most famous deafblind person, a breast exam. What are you going to do?



Helen Keller

- Systematic desensitization is often practiced covertly, that is, the person imagines the anxiety-provoking situation and then practices maintaining deep relaxation.
- Once the person feel comfortable imagining the first situation, then the second situation on the list is presented, and so forth.
- Once the person is desensitized to the entire situation list, they can begin brief exposures in the natural environment, gradually increasing the time, while continuing to remain relaxed.

Systematic Desensitization and Graduated Exposure

- Systematic desensitization is a procedure that replaces a fear response with a relaxing response in a previously anxiety-provoking situation
- Replace anxiety, fear and escape and avoidance behaviors with relaxing behaviors
- Three steps:
 1. Make a list of things in the situation that cause anxiety from the least anxiety-provoking to the most anxiety-provoking
 2. Teach a relaxation technique (often deep muscle relaxation or deep breathing)

- The desensitization can also take place in a virtual environment (video, film, or on a "set"), in the natural environment, in individual or group sessions.
- Research supports the efficacy of use of the virtual or natural environments.
- This treatment is often used by cognitive behavior therapists and other clinicians to treat irrational fears such as fear of snakes, spiders, dogs, elevators, bridges, heights, public speaking, etc.
- However, due to the cognitive requirements of "imagining" a situation and the very specific directions for deep muscle relaxation, this type of treatment may not be as helpful with developmentally delayed people.

Graduated Exposure

- Gradual systematic exposure to anxiety provoking situations in a slow step-wise manner.
- 7 Steps in graduated exposure:
 1. List behavioral expectations: what do you want the person TO DO?
 2. Make a list of things in the situation that cause anxiety from the least anxiety-provoking to the most anxiety-provoking
 3. Present an item from the situation list starting with the least anxiety-provoking to the most anxiety-provoking

- If the person is displaying behavioral difficulties, abandon the trial and look at your situation list. Do you need to change something? Was something different in the exposure? Was the step too big?
- Sometimes pairing a highly preferred activity with the exposure step (example: music, tactile feedback, lollipop, cookie bits, warm neck wrap, vibrating object) can be helpful.
- Once you have worked through the situation list, introduce new variations one at a time to increase generalization.
- New variations can include new staff, new personnel, new room, different order of steps, etc.

4. Use preferred activities during and reinforcers after the exposure to increase comfort and motivation
5. Go forward only when successful with previous exposure
6. Take the person's lead. Go only as fast as their comfort level allows. Do not force. If the person becomes even a bit anxious, stop immediately and leave.
7. This treatment also can be paired with modeling e.g. watching someone else remaining relaxed in the situation

- Take your time. Go slowly.
- Evaluate after each exposure.
- Maybe the person needs more than three successful exposures
- If possible use communication to increase comfort level
- Teach deep breathing by practice and modeling
- You can use this procedure, even with more intrusive procedures and interventions

Tips for Making Graduated Exposure In Medical Settings More Successful

- Use most preferred person to accompany the person to an exposure
- Use mood induction (Carr et al., 2003) before and during the exposure (get the person in a good mood by music, snacks, activities, etc.)
- Practice regularly (daily, weekly if you can manage it)
- Do not go beyond the step you are on because the person "is having a good day"
- Stop when your exposure is successful, reinforce and leave
- Use three consecutive successful exposures as the criterion to move to the next step

Situation List: Example

- Sit in the waiting room for 1, 3, 5, 10 minutes.
- Walk into the exam room and sit down in a chair for 15 seconds (30, 60, 120 seconds).
- Sit on the exam table for 15 seconds.
- Allow Dr. to apply stethoscope to heart and lungs for 5 seconds (then increase time systematically)
- Allow Dr. to look in one ear
- Allow Dr. to look in both ears
- Allow Dr. to take temperature
- Allow Dr. to take blood pressure

- Allow Dr. to take blood pressure:
 1. Apply cuff (no pressure), 5 seconds
 2. Apply cuff (little pressure), 5 seconds
 3. Apply cuff (medium pressure) 5 seconds
 4. Apply cuff (maximum pressure) but release pressure immediately
 5. Apply cuff (maximum pressure) but gradually increase length of time 5 seconds, 10 seconds, 15 seconds, etc.

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The best and most beautiful things in the world cannot be seen, or even touched, they must be felt with the heart.

Helen Keller

Resources: Vision

- American Council of the Blind - acb.org: advocacy and public policy group for parents
- American Foundation for the Blind - afb.org: education, resources, consultation
- American Printing House for the Blind - aph.org: nonprofit publishing house in Braille and large type formats
- Lighthouse International - lighthouse.org: information about infants and young children with visual impairments
- National Association for Parents of the Visually Impaired (NAPVI) - spedex.com: by & for parents of children with visual impairments, support groups, info and referrals
- Perkins Braille & Talking Book Library –Perkins.org/btbl/: provides free services to MA residents of any age who are unable to read traditional print materials due to a visual or physical disability
- Adaptive Technology Consulting – adaptivetech.net: demonstration center for products for blind, low vision, and learning disabled clients

Graduated Exposure Situations

- Nail cutting
- Hair cutting
- Showering (vs. bathing)
- Tooth brushing
- Blood pressure
- Blood draw
- Throat culture
- Flu shot
- Walking into the medical facility

Resources: Hearing

- National Assoc of the Deaf - nad.org: consumer advocacy organization
- Helen Keller National Center for the Deaf-Blind Youths and Adults - helenkeller.org: advocacy, info and referral
- National Deaf Education Network and Clearinghouse - Gallaudet.edu/nicd: info and referral

CHARGE Syndrome

A genetic condition: "a mutation at CHD7 on chromosome 8".
First described in 1981

The major characteristics are the 4 Cs:

- Coloboma: lesion or defect of the eye, usually a fissure or cleft of the iris, retina, optic nerve, microphthalmia
- Choanal atresia: (posterior of the nares)
- Cranial nerve dysfunction or abnormality
- CHARGE ear
- Other "minor" criterion such as delayed growth and puberty, endocrine, cardiac, renal, urinary disorders. So...the "C"s are the most important criteria here
- Behavioral and intellectual disorders are typical
- There is GREAT variability with CHARGE syndrome from some children very mildly affected in all areas and do well in public school. And.. some who are very significantly affected (total blindness, profound deafness, balance/motor, cognitive, and developmental impairments) who require very special educational services.
- Many of these children are very sick at birth and do not survive. Others face many surgeries including tracheotomy, G-tubes, heart repairs, and others.

Septo-Optic Dysplasia

A rare birth defect characterized by:

- abnormal development of the optic disk
- pituitary deficiencies
- often agenesis (absence) of the septumucidum (the part of the brain that separates the anterior horns or the lateral ventricles of the brain).
- Pituitary deficiencies (inadequate growth hormone)
- Seizures may also occur.
- Intellectual problems vary in severity among individuals. While some children with SOD have normal intelligence, others have learning disabilities and mental retardation.

Retinopathy of Prematurity

- **The major contributing factors are low birth weight and hyperoxia**
- **Their visual conditions or in some cases, total blindness, is a result of ROP or what was called Retrolental Fibroplasia (growth of abnormal blood vessels and scar tissue that can damage the retina).**
- **Often, because these babies are very premature, they also face life-threatening infections and other health conditions that require extensive medications and other invasive treatments**
- **They often have multiple cognitive and physical disabilities**

Laurence-Moon-Biedle Syndrome

- A rare inherited condition

Characteristics are:

- rod/cone dystrophy (atypical retinitis pigmentosa - a progressive eye condition which can lead to blindness)
- obesity (usually with an early onset and resistant to treatment)
- polydactyly
- hypogonitalism
- mild to severe learning difficulties
- kidney malformations and renal dysfunction
- usually four out of these six features are required to make the diagnosis, but there are other important characteristics including: developmental delay; speech difficulties; olfactory deficits; diabetes mellitus; diabetes insipidus; hepatic fibrosis; and hormonal deficiencies (e.g. thyroid, testosterone).

Usher Syndrome

- Genetic Disorder, inherited autosomal recessive
- Usher syndrome is the most common condition that affects both hearing and vision.
- The major symptoms are hearing loss and an eye disorder called retinitis pigmentosa, or RP.
- Retinitis pigmentosa causes night-blindness and a loss of peripheral vision through the progressive degeneration of the retina. As retinitis pigmentosa progresses, the field of vision narrows, a condition known as "tunnel vision," until only central vision remains.
- Balance disorders are common.

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