
Hearing and Visual Impairments in Children with Intellectual and Developmental Disabilities

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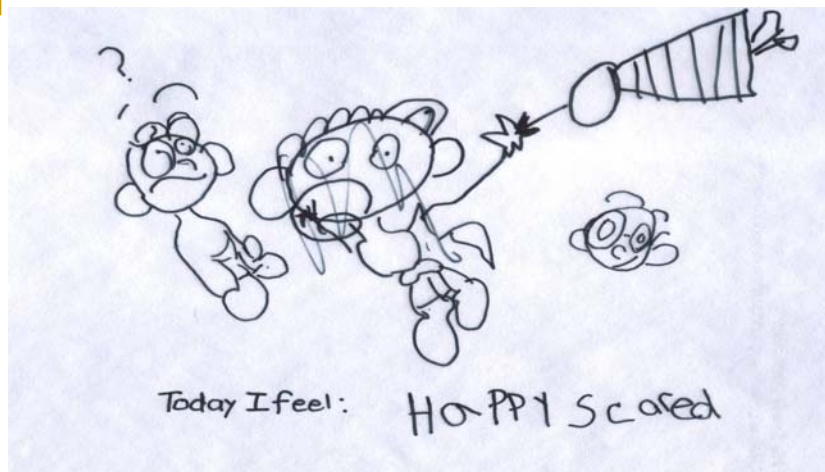
Perkins School for the Blind

Watertown, MA

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Every Picture Tells a Story



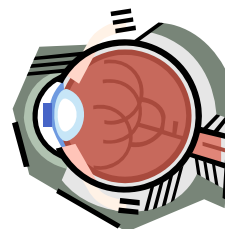
Objectives

- What does it mean to be Blind, Deaf or DeafBlind
- How best to communicate with children with sensory impairments and help their parents
- Strategies for assisting children and their parents with sensory impairments

- Children who are deaf, blind or deafblind often have associated physical, intellectual, behavioral, social, medical or motor problems
- The eye is affected in about ¼ of all inherited disorders
- Effective communication requires putting hearing and visual impairments in context with all the other problems
- Effective communication means learning the best means of communicating with each child, remember each individual is unique
- Teaching strategies should be based upon the child's particular sensory impairments and unique communication skills

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■ Visual Impairments



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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

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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
 - children with visual field loss need O&M and visual scanning training

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- 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 care be caused by the eye (cataracts) or externally; cataracts can be seen in children, not just adults

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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 began to provide benefits to the blind; the visual fields part was added the following year

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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

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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
- Difficulty eating, eye rubbing

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Vision Loss Consequences

- Delay in learning ADL's (activities of daily living)
- Restrictions in mobility without training
- Limits access to education, recreation, employment, socialization
- Environmental safety concerns
- Sense of, or actual isolation
- Depression

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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
- children 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

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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

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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

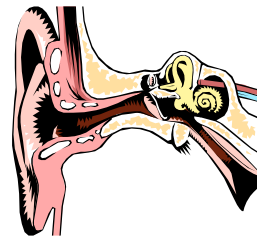
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Safety

- 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 child
- 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
- Improve lighting on stairs and closets
- Improve outdoor lighting on walkways and stairs
- Contrast on steps
- Approach the child and put items for use on the side with the better vision

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■ Hearing Impairments



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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)

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Types of Hearing Loss

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

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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

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Get an hearing evaluation if you notice **new onset of:**

- Difficulty understanding words, especially against background noise or in a crowd of children
- Needing to turn up the volume of the television, radio or iPod
- Withdrawal from conversations and social interactions
- Behavioral changes such as inattention in loud or busy settings

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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 including BAHA (bone anchored hearing aid)
- ❑ Takes time to adapt and many children 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

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Baha device

Bone Anchored Hearing Aid



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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)

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Cochlear Implant



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Additional Devices and Communication Techniques

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

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Is a Hearing Aid Appropriate and Useful For a Child Who May Not Easily Tolerate One ?

- Benefits of hearing aids: assists with communication, allows the child to enjoy music or other sounds, makes the child 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 in a small but increasing amount of time the hearing aid is tolerated

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■ DeafBlindness



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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

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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 also occur anytime birth due to meningitis, brain injury or inherited conditions*

* From National Consortium on Deaf-Blindness (2007)

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Facts About Deafblindness

- 10,000 children are deafblind
- 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

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Causes of Deafblindness

- No known etiology-17%
- Hereditary syndomes-13%
- Usher syndrome is the most common cause
- 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

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

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

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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

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- *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)

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As a Parent or Teacher of a Sensory Impaired Child You Can:

Develop a Sensory Profile

- The Profile should include:
 - Expressive communication skills and strategies
 - Receptive communication skills and strategies
 - Equipment use and non-use
- Situations you might use it include:
 - Medical appointments
 - Recreation activities
 - When starting a new day care or school
 - Other community activities

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Interactions with Children Who Have Hearing and/or Visual Impairments

- Always identify yourself immediately
- Determine the child's preferred strategies for communication
- Don't rush, give the child a chance to process the information
- Eliminate any distracting noises
- Tell the child what you plan to do and what to expect
- Do not make assumptions about the assistance the child needs, ask the parents and/or child first, offer assistance, don't just do

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Interacting with Children Who Have Visual Impairments

- Describe the environment to the child before or as entering it, guide the child's hand to chairs, desks, other furniture, sinks, toilets, toilet paper holders, etc...
- Allow the child, especially older children, to explore the room to get oriented, remember the blind child sees with his/her hands
- When walking with the child offer your elbow and walk in front of the child, describing doorways, stairs etc... just before you reach them (sighted guide)
- Touch should be firm, but gentle, light touch can be difficult to tolerate
- Never leave the child in free space, have them touch a wall, chair, etc... and let the child know before you leave the area
- Try not to move around too much so the child can stay oriented to you

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- Allow the child to explore the environment
- Described what is going on around the child, what you are doing away from the child
- Explain any written instructions (posted, handouts, signs), especially important for older children
- Keep the child informed and aware of the setting and activities using clear precise descriptions
- Do not pet or interact with guide dogs
- Provide toys and activities that are tactile and in bright primary colors
- Use real life activities such as sorting clothes or silverware
- Limit the number of toys and stimulation at one time

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Interacting with Children Who Have Hearing Impairments

- Always face the child and always talk to the child, even if there is an interpreter, have good lighting on your face
- Speak slowly and clearly
- Remain in one area while talking, don't move around or turn your head away or your back towards the child
- Seat the child close to the speaker and with a clear view of the whole group
- Allow the child to move to a better listening and viewing position as he/she wishes

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- Allow time for an interpreter, if present
- Direct communication and approach to the better ear
- Use as many visual aids as possible
- Reduce the amount of noise in the environment
- Encourage the child to let you know when he/she doesn't understand, they may sometime be reluctant to do so
- Don't expect continuous attention from the child
- They have to work harder to listen and the concentration required to hear may be tiring

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The Sensory Profile

- Describe the sensory impairment
- Preferred mode of communication, both receptively and/or 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 implant, etc.
- Tips: “special things that are helpful to me”

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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:

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Example of a Sensory Profile for a Blind Child

- I am a totally blind child 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 parent or babysitter.
- 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.

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- Please tell me what is going on around me. A verbal description can be very helpful.
- Please tell me before you touch me.
- If we are going to move from one place to another, I move through 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.
- If we are going to sit down, please place my hand on the chair so I know where to sit.
- Never leave me standing in free space.

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Example of a Sensory Profile for a Deaf Individual

- I am a totally deaf child 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.

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- Please say what is happening next before it starts. The interpreter will let me know. Give us a minute to communicate.
- If I don't have an interpreter with me, please use a written note to communicate. Gestures, modeling and pantomime also can be helpful. I will appreciate you trying to communicate with me.

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Example of a Sensory Profile for a Deafblind Child

- I am a deafblind child with severe developmental disability. I can see some light and shadow and hear loud noises. I can not hear speech. I understand simple signed English, 2-3 words at a time from my parent or respite worker who will sign into my hands. I can follow very simple one step directions.
- If you ask a simple question and wait for someone to sign to me, I can respond. I have a reliable “yes” and “no” response.
- Please tell my parent or respite worker what is happening before it starts. They will tell me. Give us **time** to communicate.

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- If you want to get my attention, it is best to touch me on the back of the hand.
- If we are going to move from one place to another, I move through 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.
- If we are going to sit down, please place my hand on the chair so I know where to sit.
- Never leave me standing in free space.

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Life as a Sensory Impaired Child

- Life can be scary
- Life can be uncertain
- There is disorientation
- There is silence
- Situations can cause anxiety, stress, fear
- Consider the following situations:
 - A blind child encountering an escalator
 - A deaf child who can not get the librarian to understand her
 - A deafblind child in a crowded area who gets jostled
 - A deaf child who want to play football but can't hear the signals
 - A multi-handicapped deafblind child who requires a blood draw and an injection during a medical appointment

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- These types of situations lead to negative experiences and refusals
- Children may refuse to go to the library, refuse to go to the Dr., refuse to play sports and maybe refuse to leave the classroom or home
- Children may display extreme behavior to avoid these unpleasant situations; yell, scream, cry, tantrum, throw items, hit teachers and parents or engage in self-injurious behavior
- One of the most common and difficult problems parents and teachers encounter with sensory impaired children

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- Adults try to calm and soothe by talking, explaining, reassuring, reasoning, promising rewards and/or threatening
- But it is difficult to change behavior with words, especially if negative behavior has resulted in avoiding these unpleasant situations in the past
- There are several solutions to these problems: graduated exposure and systematic desensitization

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Graduated Exposure

- Gradual systematic exposure to anxiety provoking situations in a slow step-wise manner.
- Can be used to prevent problems before they happen or assist with problems after they occur
- 7 Steps in graduated exposure:
 1. List behavioral expectations: What do you want the child 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.

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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 child's lead. Go only as fast as their comfort level allows. Do not force. If the child 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.

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Tips for Making Graduated Exposure More Successful

- Use most preferred person to accompany the child 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 child "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

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- If the child 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 people, new places, different order of steps, etc.

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- Take your time. Go slowly.
- Evaluate after each exposure.
- Maybe the child 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 with a wide variety of situations

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■ **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 health care provider to look in one ear
- Allow health care provider to look in both ears
- Allow health care provider to take temperature
- Allow health care provider to take blood pressure

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- Allow health care provider 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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■ **Graduated Exposure Situations**

- Going to a new school
- Going to a new classroom
- Going to a medical facility
- Going to the dentist
- Going to the physical therapist
- Wearing glasses
- Wearing hearing aids
- Playing integrated sports
- Using a public pool
- Riding the school bus
- Going to the supermarket

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Systematic Desensitization

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

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

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- Systematic desensitization is often practiced covertly, that is, the child imagines the anxiety-provoking situation and then practices maintaining deep relaxation
- Once the child feels comfortable imagining the first situation, then the second situation on the list is presented, and so forth
- Once the child 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.

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- 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 for all children

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Where to Get Help with Graduated Exposure and Systematic Desensitization

- Pediatric medical center (Children’s Hospital)
- Local mental health center
- School counselor or school nurse
- Social worker
- Behavior analyst (BCBA)/go to www.bacb.org and search by zip code to find someone near you

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Keep This Boy Smiling



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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



Thank you

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Resources: Vision, DeafBlind

- 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
- Healthyvision2010.org

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- National Eye Institute
2020 Vision Place
Bethesda, MD 20892-3655
(301) 496-5248
www.nei.nih.gov
 - DB-LINK: National Information Clearinghouse on Children Who are Deaf-Blind
NCDB : The Teaching Research Institute : 345 N. Monmouth Ave. :
Monmouth, OR 97361
Voice: 800-438-9376 | TTY: 800-854-7013 | Fax: 503-838-8150
info@nationaldb.org
- New England Center Deafblind Project, www.necdbp.org,
NEC@Perkins.org.

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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
- National Institute for the Deaf
NIDCD Information Clearinghouse
1 Communication Avenue
Bethesda, MD 20892-3456
Toll-free Voice: (800) 241-1044
Toll-free TTY: (800) 241-1055
Fax: (301) 770-8977
E-mail: nidcdinfo@nidcd.nih.gov

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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:** (abnormal formation of posterior of the nares)
- **Cranial nerve dysfunction** or abnormality
- **CHARGE** ear malformations

Other characteristics include:

- Hearing and visual impairments
- Other "minor" criterion such as delayed growth and puberty, endocrine, cardiac, renal, urinary disorders,
- Eating and swallowing disorders.
- Behavioral including OCD behaviors and intellectual disorders are typical,
- There is great variability with CHARGE syndrome. Some children are very mildly affected in all areas and do well in public school. Some are very significantly affected (total blindness, profound deafness, balance/motor, cognitive, and developmental impairments) and 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.

Reference: <http://www.chargesyndrome.org/>

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Children with CHARGE



<http://www.chargesyndrome.ca/IntroducingCHARGEbooklet.htm>

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Retinopathy of Prematurity

- The major contributing factors are low birth weight (under 2 ¾ lbs) , prematurity (under 31 weeks), and hyperoxia (high concentrations of oxygen therapy)
- 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
- There are approximately 28,000 children born under 2 ¾ lbs per year, 90% have a mild form of ROP so most do not need treatment
- 400-600 children become legally blind
- References: <http://www.nei.nih.gov/health/rop/index.asp>
<http://www.ropard.org/>

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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
- Reference: www.nei.nih.gov/health/ushers/
www.usher-syndrome.org/

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Usher Syndrome Types

<http://www.nidcd.nih.gov/health/hearing/usher.asp>

	Type 1	Type 2	Type 3
Hearing	Profound deafness in both ears from birth	Moderate to severe hearing loss from birth	Normal at birth; progressive loss in childhood or early teens
Vision	Decreased night vision before age 10	Decreased night vision begins in late childhood or teens	Varies in severity; night vision problems often begin in teens
Vestibular function (balance)	Balance problems from birth	Normal	Normal to near-normal, chance of later problems

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Septo-Optic Dysplasia

A rare birth defect characterized by:

- Abnormal development of the optic disk, blindness
- Often agenesis (absence) of the septum pellucidum (separates ventricles in the brain).
- Pituitary deficiencies including inadequate growth hormone
- Seizures may also occur.
- Intellectual problems vary in severity from normal intelligence to learning disabilities or intellectual and developmental disabilities.
- References:
 - www.magicfoundation.org/www/docs/101.115/optic-nerve-hypoplasia-septo-optic-dysplasia
 - www.ninds.nih.gov/disorders/septo_optic_dysplasia/septo_optic_dysplasia.htm

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Laurence-Moon-Bardet-Biedle Syndrome

A rare inherited condition with the following characteristics:

- 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 (extra fingers or toes)
- Under developed genital organs
- 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 (smell) deficits, diabetes mellitus; diabetes insipidus; hepatic fibrosis; and hormonal deficiencies (e.g. thyroid, testosterone).

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