



Professional Day

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An Educational Checklist for CHARGE Syndrome

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Presenter Information

Lily Slavin is a second year school psychology doctoral student at Central Michigan University. She is a member of the CHARGE Syndrome Research Lab and has been researching CHARGE Syndrome since 2016. Her research focus includes educational needs of individuals with CHARGE Syndrome and methods of addressing those needs in schools.

Presentation Abstract

The Educational Checklist for Individuals with CHARGE Syndrome was developed in collaboration with an international panel of experts, the CHARGE Syndrome Research Lab at Central Michigan University, and a sampling group of parents, professionals, and state deafblind project employees. This presentation will discuss the development of the Educational Checklist and implications for professional practice. The presentation will include an overview of the educational needs commonly experienced by individuals with CHARGE Syndrome and examples of strategies professionals can use to address those needs in the schools.

Learning Objectives

- Audience members will learn about the breadth of educational needs commonly experienced by individuals with CHARGE Syndrome.
- Audience members will learn how to address and accommodate the educational needs of individuals with CHARGE Syndrome in the schools.
- Audience members will learn how families and educators can work together as a team to improve educational outcomes for individuals with CHARGE Syndrome.

A Checklist of Educational Needs for Children with CHARGE Syndrome

The following checklist is a tool that can be used by school teams and early intervention specialists to help guide educational services for individuals with CHARGE syndrome (see Glossary for elaboration on terms). CHARGE syndrome is a rare genetic syndrome occurring in about 1:10,000 to 1:15,000 births; most cases of CHARGE syndrome are caused by mutations in the *CHD7* gene. CHARGE is an acronym, which stands for some of the common characteristics (defined in glossary) exhibited by individuals with the syndrome:

- Coloboma of the eye
- Heart defects
- Atresia of the choanae
- Restriction of growth and/or development
- Genital hypoplasia
- Ear abnormalities and deafness

Individuals with CHARGE syndrome display a wide range of CHARGE characteristics. Formal diagnosis may be based on genetic testing of the *CHD7* gene, but also by the presence of a variety of major anomalies (i.e., coloboma, choanal atresia/stenosis, cranial nerve anomalies, and a characteristic CHARGE ear) and minor anomalies (i.e., genital hypoplasia, developmental delay, heart malformations, short stature, orofacial clefting, and tracheoesophageal fistula), making CHARGE a highly variable population.

Because every individual with CHARGE syndrome exhibits a unique combination and degree of CHARGE characteristics, this checklist will need to be tailored precisely to the individual. The checklist can be tailored to the child by checking the boxes to the right of each CHARGE characteristic.

The information provided in this checklist is based on recommendations from professionals and experts in CHARGE syndrome. However, because of the complexity of CHARGE syndrome, it is impossible to make it comprehensive. Accommodating students with CHARGE syndrome requires a degree of creativity. As such, this checklist should be utilized as a starting point for brainstorming possible services for individuals with CHARGE syndrome. Recommendations must be tailored to the specific individual, and monitored for effectiveness. This checklist should be used and implemented by a multidisciplinary team.

The checklist includes five categories: (1) Characteristics and Concerns, (2) Educational/Support Needs, (3) Team Members, (4) Examples of Strategies and Accommodations, and (5) Consulting Professionals. The definition of the categories are as follows.

- Characteristics and Concerns: This category includes educationally relevant characteristics commonly displayed by individuals with CHARGE syndrome. This category also includes some of the concerns or related difficulties commonly experienced by individuals with CHARGE syndrome.
- Educational/Support Needs: This category lists educational needs that result from the characteristics and concerns. Essentially, this category provides guidance on specific considerations school staff should be aware of if the individual with CHARGE syndrome exhibits each specific characteristic/concern.
- Team Members: This category lists which professionals might be directly involved in addressing the educational needs. These professionals would likely be involved in on-going service delivery.

- Examples of Strategies and Accommodations: This category provides a limited sample of specific accommodations and strategies that can be adopted to address specific needs. The team should discuss the needs of the student and add to or modify the list in order to insure an individualized program of supports.
- Consulting Professionals: This category lists professionals outside of the school with whom it may be beneficial to consult. These consulting professionals may be important for diagnosing specific characteristics or providing guidance for treatment plans, but will not be involved in day-to-day service delivery.

The checklist also provides a glossary for elaboration on key terms, and a list of recommended resources that may be beneficial in understanding the educational needs of individuals with CHARGE syndrome.

Name of individual with CHARGE syndrome: _____

	Characteristics and Concerns	✓	Educational/Support Needs	Team Members <i>(Parents and special education teachers should be consulted in every area)</i>	Examples of Strategies and Accommodations <i>(Team should <u>discuss and modify</u> to fit individual needs)</i>	Consulting Professionals <i>(the State Deafblind Projects should be consulted in numerous areas)</i>
Medical	Heart defects		Understanding stamina/fatigue and lifting limitations	Nurse; Special Education Teacher	Provide additional travel time when walking ⁵	Primary Care Physician; Medical Specialist
	Gastrointestinal issues-abdominal pain (gas)		Movement/activity; tracking bowel movements	Nurse; Special Education Teacher	Ensure availability of restroom ⁵	Primary Care Physician; Medical Specialist
	Gastrointestinal issues-feeding issues		Scheduling and location of feeding; understanding issues with chewing and swallowing; training in mode of feeding	Nurse; Lunch Staff; Speech-Language Pathologist; Behavior Consultant; Occupational Therapist	Having someone trained in how to feed ^{5, 12}	Primary Care Physician; Medical Specialist
	Breathing difficulties		Understanding stamina and posture	Nurse; Special Education Teacher	Provide additional travel time when walking ⁵ ; availability of medication	Primary Care Physician; Medical Specialist
	Chronic pain		Recognizing pain behaviors; medication management	Nurse; Special Education Teacher; School Psychologist; Behavior Consultant; Intervener	Functional behavior assessment; interpretation of pain behavior as communication ⁵ ; availability of medication; teach how to communicate pain	Primary Care Physician; Medical Specialist
	Medication management		Up-to-date list of all medications; administration of medication	Nurse; Special Education Teacher	Availability of medication ⁵ ; training in side effects	Primary Care Physician; Medical Specialist; Psychiatrist

	Immunodeficiency		Understand frequent illnesses and absences, and need for attention to cleanliness in the school	Intervener; Nurse; Special Education Teacher	Access to school material from home/hospital; ensuring a clean school environment	N/A
Sensory	Ocular defects (vision loss)		Access to visual environment; understanding of functional vision; understanding risk of retinal detachment; equipment management for vision loss; understanding of light sensitivity; understanding of visual field	Teacher Consultant for the Visually Impaired; Certified Orientation and Mobility Specialist	Functional vision assessment ⁵ ; environmental accommodations ^{3, 4, 8, 9, 10, 12} (e.g., large print, braille, angled work surface, consideration of lighting; minimize visual clutter, etc.); glasses ⁵ ; establishment of communication bubble ^{4, 10}	Ophthalmologist; Optometrist; Low Vision Specialist; State Deafblind Projects
	Auditory issues/ear abnormalities (hearing loss)		Access to auditory environment; understanding degree of hearing loss; equipment management for hearing loss	Teacher Consultant for the Hearing Impaired	Functional hearing assessment ⁵ ; assistive technology ^{4, 12} (e.g., amplification system, hearing aid); medical interventions ^{5, 12} (e.g., implants, BAHAs); minimize auditory distractions ^{3, 10} ; sign language ¹²	Audiologist; Ear, Nose, and Throat Specialist; State Deafblind Projects
	Deafblindness		Access to environment; understanding the complexity of <u>combined</u> vision and hearing loss; knowledge of deafblind-specific intervention	Teacher of the deafblind; Intervener	Simultaneously address the impact of visual and hearing loss ^{3, 10, 12} ; make information available through multiple sensory systems ³ ; direct instruction ⁹	State Deafblind Projects; Ophthalmologist; Audiologist

Vestibular system impairment (balance)		Balance; independence of mobility; understanding need for unconventional positions (e.g., horizontal, upside down, etc.)	Physical Therapist; Occupational Therapist	Vestibular system stimulation ^{4, 5, 10, 11} (e.g., swinging, rocking); provide opportunities for movement ¹⁰ ; appropriate physical supports ^{1, 3} ; seating accommodations ^{1, 3} ; support for walking ^{1, 11}	Ear, Nose, and Throat Specialist
Vestibular system impairment (vestibulo-ocular reflex)		Stable visual environment	Teacher Consultant for the Visually Impaired	Keep objects stable ^{1, 10}	Ophthalmologist; Optometrist
Proprioceptive system impairment		Opportunities for proprioceptive input (i.e., activities that support the individual being in touch with their body); understanding availability to learn; training in joint compression and firm touch	Physical Therapist; Occupational Therapist	Proprioceptive system stimulation ^{1, 3, 5, 11} ; seating accommodations ^{1, 3} ; provide deep pressure and joint compression ^{1, 10} ; support for walking ^{1, 11}	N/A
Sensory processing issues		Managing arousal levels; understanding availability to learn	Occupational Therapist	Sensory stimulation ^{4, 5, 10, 11} ; allow the child to choose and refuse sensory experiences ¹²	State Deafblind Projects; Neuropsychologist
Touch/tactile defensiveness		Understanding touch preferences (e.g., firm touch) and how the individual utilizes touch to access information	Occupational Therapist	Allow the child to choose sensory experiences ¹² ; use firm touch ¹¹	Neuropsychologist; State Deafblind Projects

	Olfactory system impairment (smell)		Awareness of limited smell and its implications	N/A	Provide information about smells in the environment ⁵ (e.g., smoke, food)	Primary Care Physician; Medical Specialist; Ear, Nose, and Throat Specialist
Developmental	Delay in intellectual/ cognitive development		Curricular modifications; encourage exploration of and access to environment; recognize the potential of the child; concept development	School Psychologist; Teacher of the Deafblind; Intervener	Modify curriculum through accommodation, adaptation, making parallel or overlapping ^{3, 5, 9, 10} ; direct instruction of functional skills ⁹	Private Licensed Psychologist; State Deafblind Projects
	Adaptive behavior		Opportunities to gain independence	School Psychologist; Teacher of the Deafblind; Intervener	Teach functional skills as prioritized by the team ⁹	Private Licensed Psychologist; State Deafblind Projects
	Posture		Seating and any other accommodations; understanding the need for “unusual” postures/positions (e.g., side lying)	Physical Therapist; Occupational Therapist	Consider postural needs in every educational activity and setting and allow for “unusual” postures ^{3, 10, 12}	Orthopedist
	Social skills/social communication		Opportunity for social interactions; develop a positive social community	School Psychologist; Social Worker; Speech-Language Pathologist; Teacher of the Deafblind; Intervener	Circle of friends ⁶ ; social stories ⁶ ; teach social-emotional skills curriculum ^{6, 8, 12}	Private Licensed Psychologist; State Deafblind Projects
	Toileting Issues		Scheduling; awareness of neurological causes; work towards increased independence	School Psychologist; Behavior Consultant	Functional behavior assessment ⁵ ; availability of restroom; diapering accommodations	Primary Care Physician; Medical Specialist; State Deafblind Projects

	Motor delay/abnormal motor pattern		Understanding of CHARGE developmental milestones for gross motor, fine motor, and independent movement	Certified Orientation And Mobility Specialist; Physical Therapist; Occupational Therapist	Environment accessibility ¹¹ ; teaching routes ¹¹ ; cane ¹¹ ; wheelchair ¹¹ ; offer position changes ^{1, 3, 11} ; encourage environmental exploration ^{4, 11}	N/A
Communication	Receptive communication (e.g., difficulties hearing, difficulties seeing, motor issues)		Communicate to child via numerous sensory channels (e.g., spoken language, sign, pictorial cues, objects, touch, cued speech, etc.)	Speech-Language Pathologist; Sign Language Tutor; Intervener; School Psychologist	Utilize a total communication approach ^{3, 4, 8, 9, 10, 11} ; provide time to process information and respond ^{8, 9}	Private Licensed Psychologist; State Deafblind Projects
	Expressive communication (e.g., vocabulary acquisition, articulation of speech and sign, breathing difficulties)		Offer multiple opportunities/methods for communicating (e.g., gestures, sign language, augmentative strategies, etc.)	Speech-Language Pathologist; Sign Language Tutor; Intervener; School Psychologist	Utilize a total communication approach ^{3, 4, 8, 9, 10, 11} ; interpret body language and gestures as communication ^{8, 9} ; teach communication based on child's interests ¹⁰ ; create a responsive environment; offer augmentative and alternative communication devices ¹²	Private Licensed Psychologist; State Deafblind Projects

Behavioral	Physical behaviors (e.g., scratching, hair pulling, biting, self-injury)	Environmental scan/ accommodations; identification of reasons for behavior (e.g., pain, sensory, environmental, etc.); determination of importance and immediacy of intervention	School Psychologist; Behavior Consultant; Teacher of the Deafblind; Intervener	Functional behavior assessment ^{9, 10, 11} ; interpret behavior as communication ^{4, 7, 11} ; address behavior without taking away communication ^{5, 11} ; consider pain and anxiety ⁵	Private Licensed Psychologist; Primary Care Physician; Psychiatrist; State Deafblind Project
	Verbal behaviors (e.g., repetitive statements or questions, yelling, complaining)	Environmental scan/ accommodations; identification of reasons for behavior (e.g., pain, sensory, environmental, etc.); determination of importance and immediacy of intervention	School Psychologist; Behavior Consultant; Teacher of the Deafblind; Intervener	Functional behavior assessment ^{9, 10, 11} ; interpret behavior as communication ^{4, 5, 7, 11} ; address behavior without taking away communication ^{5, 11} ; consider pain and anxiety ⁵	Private Licensed Psychologist; Primary Care Physician; Psychiatrist; State Deafblind Projects
	Nonverbal behaviors (e.g., agitation, pacing, withdrawal)	Environmental scan/ accommodations; identification of reasons for behavior (e.g., pain, sensory, environmental, etc.); determination of importance and immediacy of intervention	School Psychologist; Behavior Consultant; Teacher of the Deafblind; Intervener	Functional behavior assessment ^{9, 10, 11} ; interpret behavior as communication ^{4, 7, 11} ; address behavior without taking away communication ^{5, 11} ; consider pain and anxiety ⁵	Private Licensed Psychologist; Primary Care Physician; Psychiatrist; State Deafblind Projects

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Glossary

- Atresia of the choanae- the back of the nasal passage (the choanae) is blocked by bony or membranous tissue
- Audiologist- professionals who provide care in the prevention, identification, diagnosis, and treatment of hearing, balance, and other auditory disorders
- Augmentative and alternative communication devices- devices utilized to help individuals with language impairments communicate
- Bone anchored hearing aids (BAHA)- a type of hearing aid that uses bone conduction to address conductive hearing loss, unilateral hearing loss, or mixed hearing loss
- Behavior consultant- a professional with expertise in behavior change
- Certified orientation and mobility specialist- a professional who teaches individuals with visual impairments to travel safely, confidently, and increasingly independently in their environment
- Cochlear implant- a surgically implanted device which provides a sense of sound to individuals with sensorineural hearing loss
- Coloboma- the term used to describe a part of the eye that has not completely formed
- Communication bubble- the space around an individual within which they are able to communicate
- Teacher of the Deafblind- a teacher with expertise and experience in deafblindness
- Deafblindness- a combined hearing and vision loss
- Ear nose and throat specialist- a medical professional specializing with conditions of the ear, nose, and throat
- Expressive communication- the ability to produce communication
- Frequency-modulated (FM) systems- an assistive listening device which uses radio signals to transmit speech directly to the listener's ears in noisy environments
- Functional behavior assessment- strategies to identify the underlying function of behavior
- Genital hypoplasia- underdevelopment of genitals
- Hearing aids- a device which improves hearing by amplifying sound to individuals with hearing loss
- Immunodeficiency- a weakened immune system
- Intervener- a para-professional who has specialized training in deafblindness and providing access to visual and auditory information missed because of the impact of having combined vision and hearing losses
- Neuropsychologist- a professional who specializes in understanding the relationship between the brain and behavior
- Occupational therapist- a professional who helps students engage in activities of daily living through therapy that addresses sensory and motor functions
- Ophthalmologist- a professional specializing in medical and surgical eye disease
- Optometrist- a professional who provides primary vision care
- Orofacial clefting- openings or splits in the roof of the mouth and/or lip
- Physical therapist- professionals who provide therapy to promote mobility and function of muscle systems
- Proprioceptive input- sensations from joints, muscles, and connective tissues
- Proprioceptive system- the unconscious awareness of one's muscles and joints, which sends information to the brain about body position, posture, and location

- Psychiatrist- a professional who diagnoses and treats mental disorders
- Receptive communication- the ability to understand information that is communicated to us
- Retinal detachment- an emergency when part of the eye, the retina, pulls away from supportive tissue, which may result in partial or complete loss of vision in the eye if untreated. If found quickly, reattachment and reduction of impact of the vision loss is possible
- School psychologist- a professional who addresses and supports students' academic, social, behavioral, and emotional development
- Speech-language pathologist- a professional who prevents, diagnoses, and treats speech, language, and communicative disorders
- State Deafblind Projects- the State Deafblind Projects offer consultative services for any child or young adult age birth through twenty-one years old who are suspected of having a combined vision and hearing loss. The State Deafblind Projects have a wealth of information about educating students who are deafblind and should be consulted with often
- Tactile defensiveness- a negative reaction or sensitivity to touch
- Teacher Consultant for the Hearing Impaired- a professional who assists students with deafness and hearing impairments in accessing classroom and school resources
- Teacher Consultant for the Visually Impaired- a professional who assists students with blindness and visual impairments in accessing classroom and school resources
- Tracheoesophageal fistula- abnormal connection in one or more places between the esophagus and trachea
- Vestibular system- provides information about balance and spatial orientation and responds to the position of the head in space
- Vestibulo-ocular reflex- the ability to focus on a stationary object while the head is in motion

Selected Reference List

Research books and articles

- ¹Brown, D. M. (2003). Educational and behavioral implications of missing balance sense in CHARGE syndrome. *California Deafblind Services, reSources*, 10(15), 1–3. Retrieved from <http://www.cadbs.org/newsletter/>
- ²Brown, D. M. (2004). ‘Knowing the child’- Personal passports. *California Deafblind Services, reSources*, 11(4). Retrieved from <http://www.cadbs.org/newsletter/>
- ³Brown, D. M. (2011). Deaf-Blindness, Self-regulation and Availability for Learning: Some thoughts on educating children with CHARGE syndrome. *California Deafblind Services, reSources*, 16(3), 1–7. Retrieved from <http://www.cadbs.org/newsletter/>
- ⁴Griffin H. C., Davis M. L., & Williams S.C. (2004). CHARGE syndrome: educational and technological interventions. *RE:view*, 35, 149–157.
- ⁵Hartshorne, T. S., Hefner, M. A., Davenport, S. L. H., & Thelin, J. W. (Eds.) (2011). *CHARGE Syndrome*. San Diego: Plural.
- ⁶Hartshorne, T. S., & Schmittel, M. C. (2016). Social-emotional development in children and youth who are deafblind. *American Annals of the Deaf*, 161, 444–453.
- ⁷Hartshorne, T. S., Stratton, K. K., Brown, D., Brown, S. M., & Schmittel, M. C. (2017). Behavior in CHARGE syndrome. *American Journal of Medical Genetics Part C*, 175, 431–438.
- ⁸Lewis, C., & Lowther, J. (2001). CHARGE association: Symptoms, behaviour and intervention. *Educational Psychology in Practice*, 17, 69–77. doi:10.1080/02667360120039997
- ⁹Smith, K. G., Smith, I. M., & Blake, K. (2010). CHARGE syndrome: An educators’ primer. *Education & Treatment of Children*, 33, 289–314.

Recommended Resources

CHARGE Syndrome Foundation: <https://www.chargesyndrome.org/>

National Center on DeafBlindness: <https://nationaldb.org/>

¹⁰**CHARGE Education Professional Packet:** <https://www.chargesyndrome.org/professionals/education-professional-packet/>

California DeafBlind Services Newsletter: <http://www.cadbs.org/newsletter/>

¹¹**CHARGE Management Manual for Parents:** <https://www.chargesyndrome.org/families/resources/management-manual-for-parents/>

¹²**CHARGE Information Packet for Practitioners:** <https://www.sense.org.uk/content/charge-information-pack-practitioners>

Why I am me: All about CHARGE Syndrome:

<https://www.chargesyndrome.org.au/product/why-i-am-me/>

Perkins School for the Blind: <http://www.perkinselearning.org/videos/webcast/charge-syndrome-overview>

Texas School for the Blind and Visually Impaired: www.tsbvi.edu

Services for students who are deafblind:

https://www.cmich.edu/colleges/class/Centers/DBCentral/Documents/Comparison_of_Supports_6.22.16.pdf