A Checklist of Educational Needs for Individuals 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.

Overview of CHARGE Syndrome

CHARGE syndrome is a rare genetic syndrome occurring in about 1:10,000 to 1:15,000 births. CHARGE is a diagnosis made by a medical geneticist based on major (i.e., coloboma, choanal atresia/stenosis, cranial nerve anomalies, and a characteristic CHARGE ear) and minor (i.e., genital hypoplasia, developmental delay, heart malformations, short stature, orofacial clefting, and tracheoesophageal fistula) anomalies and genetic testing (*CHD7* gene mutation). Please see the selected reference list at the end for more information.

CHARGE syndrome is extremely complex and variable. School teams must understand these five domains in which individuals with CHARGE are commonly affected:

1. **Medical**: Individuals with CHARGE syndrome are extremely medically complex. They often spend the first several years of their lives in and out of the hospital undergoing numerous life-threatening surgeries. Educational teams must understand the extent of the medical complexities individuals with CHARGE experience and that medical needs will take priority over educational needs.
2. **Sensory**: Every sensory system can be affected by CHARGE syndrome: vision, hearing, taste, smell, touch, proprioception, and vestibular. A vast majority of individuals with CHARGE experience ear abnormalities and deafness, coloboma of the eye and vision loss, and vestibular abnormalities, making it difficult for them to access information from their environment. Educational teams need to recognize and address the sensory loss of individuals with CHARGE to maximize the sensory input the individual receives.
3. **Communication**: Due to multiple sensory impairments and structural anomalies (e.g., cleft lip, choanal atresia), individuals with CHARGE syndrome often have difficulties understanding and communicating with others. As such, they often communicate the only way they can: their behavior. Educational teams will need to learn to recognize behavior as communication and find creative ways to expand the expressive and receptive communication of individuals with CHARGE syndrome. Individuals communicate in a variety of ways (e.g., spoken, sign, gestural, behavioral, pictorial, etc.), so establishing a communication system that works for the individual is a priority.
4. **Developmental**: Global developmental delays are universal in CHARGE syndrome due to individuals' medical fragility, multiple sensory impairments, and physical anomalies. However, these delays can be addressed in the school by a variety of educational team members.
5. **Behavioral**: Due to multiple sensory impairments and subsequent difficulties with communication, individuals with CHARGE syndrome often initially communicate with their behavior. A CHARGE behavioral phenotype has been suggested, which includes the following characteristics: 1) low normal cognitive functioning, 2) socially interested but immature, 3) repetitive behaviors, which increase under stress, 4) high degree of sensation seeking, 5) tendency to lose behavioral control and self-regulation when stressed or sensory overloaded,

and 6) difficulty shifting attention. Sources of problem behavior are believed to be pain, sensory issues, and anxiety.

Overview of the Checklist

This tool is comprised of the following components: introduction (overview of CHARGE syndrome and checklist), checklist, glossary, and recommended resources. The information provided in this checklist is based on recommendations from experts in CHARGE syndrome. However, because of the complexity of CHARGE syndrome, it is impossible to make it exhaustive. Accommodating students with CHARGE syndrome requires a degree of creativity. As such, this checklist should be utilized by a multidisciplinary team 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.

The checklist includes five categories:

1. **Characteristics and Concerns**: Educationally relevant characteristics and concerns/related difficulties commonly displayed by individuals with CHARGE syndrome. Check the box to the right of each CHARGE characteristic exhibited by the individual.
2. **Educational/Support Needs:** Educational needs that result from the characteristics and concerns. These are specific considerations school staff should be aware of if the individual exhibits each specific characteristic/concern.
3. **Team Members:** Professionals who might be directly involved in addressing the educational needs via on-going service delivery.
4. **Examples of Strategies and Accommodations:** Limited sample of specific accommodations and strategies that may 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.
5. **Consulting Professionals:** Professionals outside of the school with whom it may be beneficial to consult regarding diagnoses or guidance for treatment plans. These will likely not be involved in day-to-day service delivery.

Name of Individual with CHARGE Syndrome: _____

	Characteristics and Concerns	✓	**Educational/ Support Needs**	**Team Members** ***(Parents, Special and General Educators, & Interveners should consulted in most areas)***	**<u>Examples</u> of Strategies and Accommodations** **(*Team should <u>discuss and modify</u> to fit individual needs*)**	**Consulting Professionals** ***(the State Deafblind Projects should be consulted in most areas)***
Medical	Heart defects		Understanding stamina/fatigue and lifting limitations	Nurse	Provide additional travel time when walking[5]	Primary Care Physician; Medical Specialist
	Gastrointestinal issues- abdominal pain (gas)		Movement/activity; tracking bowel movements	Nurse	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	Feeding therapy; cut foods into small bites; train staff on feeding[5, 12]	Primary Care Physician; Medical Specialist; Dietician
	Breathing difficulties		Understanding stamina and posture	Nurse	Provide additional travel time when walking[5]; medication management	Primary Care Physician; Medical Specialist
	Chronic pain		Recognizing pain behaviors; medication management	Nurse; School Psychologist; Behavior Consultant	Functional behavior assessment; interpretation of pain behavior as communication[5]; medication management; teach how to communicate pain	Primary Care Physician; Medical Specialist

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Medical	Medication management		Up-to-date list of all medications; administration of medication	Nurse	Availability of medication[5]; training in side effects	Primary Care Physician; Medical Specialist; Psychiatrist
	Immuno-deficiency		Understand frequent illnesses and absences, and need for attention to cleanliness in the school	Nurse	Access to school material from home/hospital; ensuring a clean school environment	Immunologist
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]; learning media assessment, 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

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Sensory	Auditory issues/ear abnormalities (hearing loss)		Access to auditory environment; understanding degree and type 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; Otolaryngologist; State Deafblind Projects
	Deafblindness		Access to environment; understanding the complexity of combined 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

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Sensory	Vestibular system impairment (balance)		Balance; independence of mobility; understanding need for unconventional positions (e.g., horizonal, 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]	Otolaryngologist
	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 his or her 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

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Sensory	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	Intervener	Provide information about smells in the environment[5] (e.g., smoke, food)	Primary Care Physician; Medical Specialist; Otolaryngologist
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	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

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Developmental	Adaptive behavior		Opportunities to gain independence	School Psychologist; Teacher of the Deafblind	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;	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	Nurse; School Psychologist; Behavior Consultant	Functional behavior assessment[5]; availability of restroom; diapering accommodations	Primary Care Physician; Medical Specialist; State Deafblind Projects; Urologist

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Developmental	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

	Characteristics and Concerns	✓	Educational/ Support Needs	Team Members *(Parents, Special and General Educators, & Interveners should consulted in most areas)*	**Examples** of Strategies and Accommodations (*Team should discuss and modify to fit individual needs*)	Consulting Professionals *(the State Deafblind Projects should be consulted often)*
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	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	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

	Characteristics and Concerns	✓	Educational/ Support Needs	Team Members *(Parents, Special and General Educators, & Interveners should consulted in most areas)*	**Examples** of Strategies and Accommodations (*Team should **discuss and modify** to fit individual needs*)	Consulting Professionals *(the State Deafblind Projects should be consulted often)*
Behavioral	Nonverbal behaviors (e.g., anxiety, 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	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- a professional who provides care in the prevention, identification, diagnosis, and treatment of hearing, balance, and other auditory disorders
- Augmentative and alternative communication (AAC) 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 (O&M) specialist- a professional who teaches individuals with visual impairments to travel safely, confidently, and increasingly independently in their environment
- Cochlear implant (CI)- 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; typically results in visual field loss
- Communication bubble- the space around an individual within which they are able to communicate
- Deafblindness- combination of at least some degree of hearing loss and some degree of vision loss
- Dietician- a professional who specializes in diet and nutrition
- Expressive communication- the ability to produce communication
- Frequency-modulated (FM) system- 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 reason behind behavior
- Genital hypoplasia- underdevelopment of genitals
- Hearing aid- a device which improves hearing by amplifying sound to individuals with conductive 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
- Learning media assessment- an assessment conducted to provide information regarding which senses an individual uses to gather information from his or her environment
- Neuropsychologist- a professional who specializes in understanding the relationship between the brain and behavior
- Occupational therapist (OT)- 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 (cleft palate) and/or lip (cleft lip)
- Otolaryngologist- a medical professional specializing with conditions of the ear, nose, and throat

- Physical therapist (PT)- a professional who provides therapy to promote mobility and function of muscle systems
- Proprioceptive input- sensations from joints, muscles, and connective tissues which help you know where you are in space
- 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 medical professional who diagnoses and treats mental disorders
- Receptive communication- the ability to understand information that is communicated to an individual
- 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 (SLP)- 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 in the United States. 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 (TCHI)- a professional who assists students with deafness and hearing impairments in accessing classroom and school resources
- Teacher Consultant for the Visually Impaired (TCVI)- a professional who assists students with blindness and visual impairments in accessing classroom and school resources
- Teacher of the Deafblind- a teacher with expertise and experience in deafblindness
- Tracheoesophageal fistula (TEF)- connection between the esophagus and trachea
- Vestibular system- structures of the inner ear (semicircular canals) which provide information about balance and spatial orientation and responds to the position of the head in space
- Vestibulo-ocular reflex (VOR)- the ability to focus on a stationary object while the head is in motion

Selected Reference List

Research Books and Articles

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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/for-professionals/education-professional-packet/

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

[11]CHARGE Management Manual for Parents: https://www.chargesyndrome.org/for-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/store/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