

# *CHARGE Syndrome Foundation, Inc.*

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## *Introduction to the Professional Packet*

Welcome to the world of CHARGE! As you have already determined, CHARGE syndrome is a very complex condition. Every child is different. Over the years, we have received inquiries from professionals who are, or soon will be, working with an individual with CHARGE syndrome. These professionals are looking for information about CHARGE that is helpful to them. There now exists a large body of information on CHARGE available to physicians caring for the medical issues in individuals with CHARGE syndrome. A gene for CHARGE (CHD7) was identified in 2004. If you are interested in more detailed medical information, see the comprehensive summary at [www.genetests.org](http://www.genetests.org) [choose the "CHARGE syndrome" article in the GeneReviews section]. The CHARGE Syndrome Foundation also publishes the **Management Manual for Parents**, which covers medical information and some developmental information on CHARGE which we have brought together. The Manual is available in .pdf format at the Foundation website ([www.chargesyndrome.org](http://www.chargesyndrome.org) at the "Resources" link) and in print from the Foundation. All of the articles from the American Journal of Medical Genetics special issue on CHARGE are also available through the Foundation website.

Although medical information is plentiful, we are only beginning to bring together the accumulating knowledge for educational and other specialists who work with individuals with CHARGE. This packet is an attempt to provide you with a brief introduction to CHARGE, its complexities, and some strategies to consider in educational and therapeutic settings. Start with the reprint of the article from **ASHA Leader**, which includes an overview of genetics, clinical diagnosis, ears and hearing, feeding, behavior and education. Then read David Brown's article on **Behavior** in CHARGE. Finally, move on to other sections in this packet, which include tips on how to think about the child with CHARGE and the classroom.

This is a work in progress. We know there are more resources out there that we don't know about. And there are lots of gaps in the knowledge that we are working to fill. Please send us feedback: let us know what parts of this were particularly useful, let us know what you would like to have available, let us know if you have come across other helpful materials. Send your feedback to the Foundation ([marion@chargesyndrome.org](mailto:marion@chargesyndrome.org)) or directly to me ([meg@chargesyndrome.org](mailto:meg@chargesyndrome.org)).

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# **CHARGE Syndrome Professional Packet**

## **Table of Contents**

Introduction	Meg Hefner
1. ASHA Leader Article	James Thelin, Editor
2. Behavior in CHARGE	David Brown
3. Welcoming the Child in the Classroom	Laurie Denno
4. Checklist for Behaviors	Martha Majors and Pam Ryan
5. Physical Characteristics	Maryann M. Girardi
6. Occupational Therapy	Anna Shinkfield
7. Speech and Language Therapy	Lisa Anne Guerra
8. Technical Assistance	Betsy McGinnity
9. Vision	Donna Bent
10. Oral Motor and Feeding	Anna Shinkfield and Lisa Anne Guerra
11. Educational Needs	Martha M. Majors and Sharon Stelzer
12. Resources for Teachers	Betsy McGinnity

## CHARGE Syndrome: Multiple Congenital Anomalies Including Disorders of All Senses and Speech, Language, Feeding, Swallowing, and Behavior

***Ears and Hearing*** by James W. Thelin and Lori A. Swanson

CHARGE syndrome is a genetic disorder (one in 10,000 to 15,000 live births) with multiple physical, sensory, and behavioral anomalies. Children with CHARGE typically undergo 10 surgeries before age 3. Although early mortality rates have been 10% to 20%, survival rates have improved with cardiac treatment. Individuals with CHARGE may have deficits in every sensory modality and frequently exhibit challenging behaviors. CHARGE is presently the leading genetic cause of deaf-blindness at birth in the United States.

For those children who survive the challenges of early life, parents focus their concerns on development, communication, and education. The ultimate level of functioning is significantly enhanced by early intervention from audiologists and speech-language pathologists. In children with CHARGE syndrome, the value of services offered by SLPs and audiologists is enhanced when other aspects of the disorder, such as clinical and genetic diagnosis, feeding and swallowing, behavior, and education, are understood. Information on behavior and development appeared in a 2004 special CHARGE syndrome edition of the *American Journal of Medical Genetics*. [available at [www.chargesyndrome.org](http://www.chargesyndrome.org) Resources Tab]



### Ears and Hearing

In CHARGE, every part of the auditory system may be involved. External ear anomalies are so distinctive that, at times, it is possible to make a preliminary diagnosis of the syndrome on the basis of pinna shape alone. (See photos above right and page 7, right.) Middle-ear ossicular anomalies can cause conductive losses as great as 70 dB and chronic otitis media secondary to eustachian tube dysfunction is nearly universal. Underdevelopment of the cochlear and vestibular structures (including Mondini's dysplasia) is common and often causes sensory losses in hearing and balance. Auditory nerve diameter may be reduced; latencies may be prolonged on auditory brainstem response tests; and agenesis of the corpus callosum has been reported. Typical losses are mixed losses with very large conductive components and substantial cochlear involvement that is usually greatest in the high frequencies. The prevalence of severe-profound hearing loss is approximately 50%.

Audiologic evaluation of children with CHARGE is challenging for several reasons:

- 1) Many children do not speak or sign.
- 2) Visual problems interfere with sound field audiometry.
- 3) Tactile defensiveness is common.
- 4) Hearing losses are often large and asymmetrical.
- 5) There may be resistance to and risks associated with sedation.

Aural habilitation also can be made difficult by: soft pinnas that do not support hearing aids well, large hearing losses that require tightly fitting earmolds to prevent feedback, and stenosed ear canals with drainage from chronic otitis media. Selecting and fitting appropriate amplification is often difficult. Some individuals have benefitted from cochlear implants and others from bone-anchored hearing aids.

Despite these challenges associated with multiple physical and sensory deficits, many children with CHARGE who are significantly involved have learned to communicate using symbolic language. Those with the greatest success have had consistent and innovative audiologic intervention very early in life and communication training (spoken and signed language)-even when the child's health was poor and hospitalizations were frequent.

### **Communication and Related Issues**

About 60% of children with CHARGE acquire symbolic language and communicate with spoken language, signs, and/or visual symbols. The mechanics of speech may be affected by craniofacial anomalies, breathing problems, and clefts. Success in acquiring symbolic language is related to communication training begun before age 3, success in overcoming hearing loss, and the ability to walk independently. One explanation for this latter finding is that an ambulatory child has the ability to move into his or her own communication bubble-the space in which the child can see and hear a communication partner optimally. Among children who use symbolic language, however, speech and language problems are common. Children who use symbolic forms often have problems in maintaining a topic and in effective turn-taking.

Children who do not acquire symbolic language may learn to demonstrate higher forms of prelinguistic communication such as use of gestures and vocalizations to regulate the behavior of others. Those who are most impaired may produce pre-intentional behaviors, which parents and caregivers may interpret as intentional. Since children with CHARGE often demonstrate a high rate of repetitive behaviors, parents and caregivers may have difficulty assigning meaning to potential communication acts.

Adapted Prelinguistic Milieu Teaching (PMT, Warren & Yoder, 2002) is currently being tested as a means to increase the rate and variety of prelinguistic communications in the deaf-blind population by Bashinski and Brady at the University of Kansas. PMT focuses on increasing use of gestural forms (e.g., distal points, gives, shows, and leading gestures), increasing rate of communication, and improving parent responsivity, which appear to be predictors of increased communication skills in children with disabilities (Brady, Marquis, Fleming, & McLean, 2004). Gestures may have some advantages over symbolic forms of communication. Unlike signs, they are readily understood by most communication partners. Most importantly, gestures may facilitate understanding of the give and take between people that underlies communication.

Children with CHARGE often also have feeding and swallowing difficulties, behavior problems unique to the syndrome, and very special educational needs. Before CHARGE was recognized as a unique disorder, individuals with the disorder were believed to have a collection of unrelated anomalies. They received treatment from many specialists that was not coordinated. Now that CHARGE is recognized, it is clear that optimal treatment results from the collaboration of specialists in medicine, communication, behavior, and education.

Problems related to CHARGE often are inter-related, and communication is an essential part of an effective clinical response. As an example, investigators at the University of Tennessee asked parents how the communication of their child with CHARGE was affected by their child's behavior. The parents responded that in many cases an inability to communicate was the cause of inappropriate behavior.

Emerging evidence suggests that early intervention by speech-language pathologists, audiologists, and educators of the deaf can enhance the acquisition of symbolic language that is crucial to communication development, social interaction, and learning. Early and persistent intervention for speech, language, swallowing, and hearing disorders can greatly enhance the quality of life for children with CHARGE syndrome-even for those who have frequent illness and those who are severely involved. An increased awareness of CHARGE syndrome should lead to enhanced services provided by professionals in our field.

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# Genetic Diagnosis

by Conny van Ravenswaaij

Although CHARGE syndrome is usually sporadic, there always has been evidence that it is caused by a genetic factor. The concordance rate in monozygotic twins is very high and affected sibling pairs do occur. We found a very small deletion of chromosome 8 in two children with CHARGE. This tiny region on chromosome 8 appeared to harbor a new gene: CHD7. Mutations in this gene were found in 16 of 17 well-selected patients. At present large groups of patients suspected for CHARGE syndrome are being studied and a mutation in CHD7 is found in about 60% of these patients. In the patients who fulfill the diagnostic clinical criteria, the mutation detection rate is as high as 90%. However, mutations can be found in patients who do not fulfill these criteria, and the mild end of the clinical spectrum is not yet known.

CHARGE syndrome has a recurrence risk of 1-2%. This can be explained by germ line mosaicism. That means that although the parent is not affected, some of his/her sperm or egg cells carry the same CHD7 mutation. Sometimes, in affected siblings, a mosaic mutation of CHD7 can be detected in one of the parents. In this situation the recurrence risk is increased. Therefore DNA studies are recommended in all parents of children with a CHD7 mutation. Prenatal diagnosis with 100% reliability is possible with chorionic villi sampling. A person with CHARGE syndrome has a risk of 50% to pass on the CHD7 mutation. However, infertility is very common in CHARGE syndrome and as a result parent-to-child transmission is rare. Further studies are in progress to determine if genes other than CHD7 can cause CHARGE syndrome.

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# ***Clinical Diagnosis***

*by Meg Hefner and Sandra L. H. Davenport*

Although CHARGE syndrome is a genetic condition most often caused by mutations in the CHD7 gene (see Genetic Diagnosis, p. 40), the diagnosis is still clinical, based on the presence of specific major and minor characteristics. Beyond the major and minor diagnostic characteristics are many additional findings in CHARGE that may be critically important in management.

Major characteristics are ocular coloboma, choanal atresia, cranial nerve abnormalities (anosmia, facial palsy, severe suck and swallow incoordination), and characteristic ears (outer ear shape, ossicular anomalies and eustachian tube dysfunction, cochlear and/or semicircular canal abnormalities). The most common features in CHARGE are cochlear and semicircular canal anomalies, which are found in >90% of children with CHARGE when imaged by MRI. Minor characteristics are those which are common in CHARGE syndrome, but are either less specific to CHARGE (e.g., heart defects), or more difficult to evaluate consistently (e.g., characteristic CHARGE syndrome face). Minor characteristics are congenital heart defects, genital hypoplasia, renal anomalies, tracheo-esophageal fistula/esophageal atresia, cleft lip or palate, growth retardation, and characteristic face and hands.

Occasional findings in CHARGE syndrome are features which may not be diagnostic by themselves, but which support a diagnosis of CHARGE and may be important in management. These include DiGeorge sequence in the absence of 22q11.2 deletion, omphalocele or umbilical hernia, bony scoliosis or hemivertebrae and additional dysmorphic features (thumb or limb anomalies, short webbed neck with sloping shoulders, nipple anomalies). If a diagnosis of CHARGE syndrome is being considered in an older individual, the typical CHARGE behavioral profile may be helpful as well. Many other syndromes (especially 22q deletion syndrome and some chromosome abnormalities) have features which overlap with CHARGE. Diagnosis of CHARGE syndrome should be made by a medical geneticist familiar with CHARGE.

Children with CHARGE syndrome show marked delays in motor development due to vestibular dysfunction in combination with prolonged hospitalization, truncal hypotonia with ligamentous laxity, decreased visual acuity and hearing impairment. Many children are reluctant to crawl, often moving by scooting in a combat crawl, pushing with their feet in the supine position or using a five-point crawl (head down). The average age of walking is 3-4 years. Language development is delayed due to multiple sensory deficits, motor deficits, and delay in establishment of an appropriate communication system.

Although developmental delays might be expected in children with CHARGE syndrome, the incidence of cognitive impairment in CHARGE syndrome is probably 50% or less. Factors which correlate with better apparent development in the teen years include age of walking and degree of visual impairment. Some children with CHARGE syndrome have specific learning disabilities such as difficulty understanding math. Many adults with CHARGE syndrome live independently and several are college graduates.

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## ***Feeding and Swallowing***

*by Sara Rosenfeld-Johnson*

Feeding problems are common in individuals with CHARGE syndrome; the causes and severity of the feeding difficulties vary greatly. The most severe of the disorders is, as the medical diagnosis states, "dysphagia: not approved for oral feedings." In this case the child could not receive any liquids or foods by mouth. Swallow studies of individuals with this diagnosis have identified physiological deficits in the swallowing mechanism. Thus, food or liquid intake can result in choking, gagging, and/or food aspiration. These individuals cannot receive food or liquid by mouth until medical approval is given.

The medical diagnosis for the second group is "dysphagia: approved for only specified food or liquid textures." Swallow studies have identified, based on the physiological deficits in the swallowing mechanism, safe and unsafe food and liquid textures. These individuals will have a diet prescribed by a speech-language pathologist trained in feeding and swallowing therapy. Changes in food textures cannot be implemented without direct communication with the physician.

The third group consists of individuals with muscle deficits secondary to weakness and/or motor-planning disorders. The swallow mechanism is intact in this group, but the preparation of the bolus for swallowing is impaired. Weakness in the muscles of the jaw, lips and tongue is often seen.

There is an additional overlying problem seen in nearly all individuals with CHARGE syndrome: sensory deficits. A complete oral sensory examination will determine the presence of oral hyposensitivity, oral hypersensitivity, mixed oral sensitivity, or fluctuating oral sensitivity. Tactile defensiveness, a fear-response to the presence or placement of certain textures within the mouth, may be superimposed over any of these.

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# ***Behavioral Phenotype***

*by Timothy S. Hartshorne*

Children with CHARGE develop behavioral difficulties, some of which may be described as autistic-like, and obsessive-compulsive, with attention difficulties and tic disorders also present. There are potentially multiple sources for these difficulties, and research is attempting to sort them out. Multi-sensory impairments, communication frustrations, and physical pain and discomfort all have been implicated. However, some children with fairly good sensory abilities, adequate communication, and little apparent discomfort may still have challenging behavior. Cognitive impairment has been implicated in some but not all cases. It seems likely that some neuropsychological problems exist. Recent research supports the presence of executive dysfunction, or problems with shifting, initiating, inhibiting, or sustaining actions based on prefrontal cortex activity.

Another area being explored is the presence of a regulatory disorder making it difficult for the child to regulate complex processes such as their sleep-wake cycle, hunger-satiety cycle, their ability to console themselves, to manage their emotions, and to plan their motor activities. Recent research has found indications of significant sleep disorders in well over half of these children, which can have a significant impact on behavior. The presence of significant stress and perhaps Post-Traumatic Stress Disorder due to pain, illness, multiple surgeries, and difficulties experienced in school and sometimes at home is also being studied. Several researchers have found that the age the child first walks is a predictor for behavioral and communication difficulties. Most children with CHARGE walk by about 3 years of age, but those who walk later are at more risk for difficulties. The behavioral difficulties are frequently managed by medication, and a wide variety of drugs have been used. Children with CHARGE may have paradoxical reactions to medications, and frequently the drugs gradually lose their effectiveness. Well-developed behavioral interventions based on functional analysis of the behavior and involving functional communication training are important.

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# Education

*by Kathleen Stremel*

Evidence has been accumulating that children with CHARGE derive the greatest benefit from early intervention and education programs that address functional hearing and vision simultaneously. Spoken and signed language together may benefit children with auditory and visual deficits. Children who are totally blind may need habilitation techniques that include tactile (e.g., touch cues, tactile sign language), movement, and sensorimotor strategies. Current early intervention and educational placements for children with CHARGE include a range of educational settings, including residential schools, separate schools, home schooling, separate classes, and regular classes. Perkins School for the Blind has the only classroom specifically for children with CHARGE.



A coordinated effort among families, early intervention specialists, and state deaf-blind projects must be a priority if children with CHARGE are to receive well-coordinated services, appropriate adaptations to access their environments, early communication systems, and intervention within the context of daily routines in natural environments. Educators in programs that serve these children are currently focusing on the following issues: the effects of multiple disabilities on educational outcomes, educational expectations with cochlear implantation, and pragmatics in communication. There is a great need for appropriate curricula for children with CHARGE, as well as parent training materials and qualified service providers.

The National Technical Assistance Consortium (NTAC) maintains the national census of individuals with deaf-blindness and provides technical assistance to deaf-blind projects in all states. These services will be continued by a new organization called the National Technical Assistance and Dissemination Center for Children and Youth Who are Deaf-Blind. The Web site for this new organization describes how the addresses for all state deaf-blind projects can be accessed. Children with CHARGE should be registered with their state deaf-blind project, which provides technical assistance to families and service providers for appropriate educational programming and other activities.

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*Invited Comment***CHARGE Syndrome “Behaviors”: Challenges or Adaptations?**

Children with CHARGE syndrome are truly “multi-sensory impaired,” having difficulties not only with vision and hearing but also with the senses that perceive balance, touch, temperature, pain, pressure, and smell, as well as problems with breathing and swallowing, eating and drinking, digestion, and temperature control. Children with CHARGE present a unique array of behaviors that are frequently reported as “challenging” [Hartshorne and Cypher, 2004], and it is true that behaviors in this population can be extreme, persistent, and apparently paradoxical.

Several decades of observing and working with children with CHARGE has shown that sometimes the behaviors that are reported as “challenging” are, in fact, adaptive responses to severe levels of multi-sensory impairment, responses that help the children to function effectively [Salem-Hartshorne and Jacob, 2004]. A therapy/educational approach that accepts and acknowledges many of these behaviors as achievements, and then uses them to work towards further skill development, trying to reduce stress levels and helping the children to develop acceptable strategies for adapting to their sensory experiences, will be more successful than one that aims, as a first priority, to remove these behaviors and replace them with more “normal” functioning [Moss, 1993].

It is important to avoid too narrow an emphasis on the “deaf-blind” aspects of CHARGE and instead consider the impact of other sensory deficits. By considering children with CHARGE from a truly multi-sensory perspective, some of the puzzling and concerning aspects of what the children do begin to seem totally explicable. Even so, many challenging behaviors are encountered that cannot be explained by this sensory perspective alone, and other factors like pain, health issues, and the impact of specific brain malformations seem to provide useful avenues for investigation. I discuss here first implications related to balance, vision, and hearing. I follow this with some comments about sensory integration (SI) and communication, and finally some anecdotes to illustrate these issues.

It is thought that most people with CHARGE have little or no balance sense due to the malfunctioning or absence of the semicircular canals (the receptors of the balance sense) in the inner ears [Admiraal et al., 1998], and to anomalies of the auditory nerve (cranial nerve VIII) [Davenport, 1999]. The semicircular canals play a crucial role in organizing sensory perception through all the other sensory channels [Murofushi et al., 1997; Maynard, 2001], and so this anomaly has a profound affect on all areas of functioning and behavior for the entire life of the child. However, its importance and impact is usually over-looked and under-played, especially once the child is standing and walking independently. Table I lists many of the effects of these balance difficulties on the young child.

Significant problems with the balance sense will inhibit the development of effective body language, since postural control, equilibrium, muscle tone, and motor coordination will all be impacted [Abadie et al., 2000]. An absent balance sense is also likely to have a negative impact on the development of memory, the effective use of vision (especially fine central vision), and the processing of auditory input, all of which have a cumulative impact on speech and language development [Colby Trott et al., 1993]. Resultant difficulties with expressing themselves, or the constant experience of having their expressive communications misinterpreted, can lead some children to give up, or to resort to explosive behaviors that may be construed as unpredictable, irrational, or excessively labile.

In later childhood and adolescence, the problems with fatigue, postural control, and sitting or standing unsupported may be less evident but still present. Sometimes the student will benefit from using an adapted chair, with arms and a footrest, possibly also with a tilted seat to encourage more active sitting. There may still be a great need to rest the head on one or both arms or even down on the desktop itself, in order to read or write. Some older children and teenagers can seem to function quite well at their desk for extended periods of time, but they then need periodically to get into a horizontal position to relax and to re-charge their energy levels for the next exertions. They may also need these periods in the horizontal position to reorganize their sensory system using behaviors like leg kicking, arm waving, shoulder shrugging, hyperventilating, or gazing at bright light [Colby Trott et al., 1993]. Extended periods standing still and entirely unsupported are usually particularly challenging.

Very persistent low muscle tone (into adulthood) is partly a complication of severe balance problems. It is also associated with low vision, breathing difficulties, and generally reduced sensory inputs, hence reduced perceptual awareness. The problem is then compounded by the lack of motivation to move and the resulting lack of “exercise.” Saving reactions, standing, cruising, and independent walking usually develop very late, as in a British survey that found a mean age of 4 years for independent walking [Blake and Brown, 1993]. When children do walk, there is often a characteristic gait, some aspects of which may remain evident for many years—feet spaced widely apart, knees bent to lower the center of gravity, body rolling from side to side with each step, feet sliding along the floor or planted down very firmly on the floor with each step (maybe several times, almost like patting the floor with the foot), and arms held up like a tightrope walker. Some children walk with repeating swaying circular movements of the upper body and head, as if trying to maintain awareness of the danger areas at the limits of safe posture.

On-going monitoring by a Physical Therapist is important because there is a high risk of the development of neuromuscular scoliosis (curvature of the spine) in childhood and the teenage years. It is important for orthopedists and therapists to recognize the neuromuscular (not bony) nature of the scoliosis because treatment is different. Delayed awareness and control of bladder and bowel movements may be attributed to poor nerve feedback due, in part, to very low tone. There appears to be no correlation between delayed toileting skills and developmental level or potential, however.

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TABLE I. Early Effects of Poor Balance Sense in Children With CHARGE

- 
- Very persistent low muscle tone (“floppy muscles”)
  - Poor head control and a generally poor ability to resist against gravity
  - Strong postural insecurity when held upright or sitting on a lap
  - A marked preference for lying flat on the back (supine), or on the side for long periods of time for most activities, including locomotion
  - Delayed mobility, then unique movement patterns including shuffling backwards head-first in supine, shuffling sideways in supine, 5-point crawling (using both knees, both arms and the forehead down on the floor for support)
  - Very persistent floor sitting with the legs in a “W” position to give a broader, more secure base
  - Certain levels of visual, auditory, communication, and fine motor skills developed while in supine position that almost all disappear (and need to be relearned) when the child is held upright
  - Bilateral coordination may be affected, with hand dominance so strong that the other side of the body may be ignored, or hand dominance may not take place at all, and eye dominance may be absent also
  - Fatigue after trying to resist gravity (e.g., by unsupported sitting or by holding the head erect) for relatively short periods of time
- 

Low muscle tone is also associated with poorly modulated tactile and proprioceptive senses, so that tactile defensiveness may be present, and awareness of touch, pain, and temperature may be fluctuating. Children often adopt specific postures (e.g., horizontal with both legs bent and one ankle up crossing the other knee, or legs tightly crossed, or fingers crossed or bunched together, or hands fistled, or arms folded). These postures provide essential extra tactile and pressure information to the brain about where the child’s limbs are in space, and also confirms for them that they are securely “fixed” and not moving or floating around. Paradoxically, after all that flat foot slapping on the floor, some children, once walking is mastered, develop a tip-toe barefoot style, the bare feet maximizing tactile input, and being on tip-toe maximizing the proprioceptive (pressure) input through the feet, ankles, calves, knees, thighs, and buttocks.

One final consideration of low tone with poor tactile and proprioceptive feedback is the need this may impose on the child to use excessive force to function, and thereby adopting a very high muscle tone, using strong movements, an over-firm grip, and excessive force in making contact with people or objects, all of which may be interpreted as aggressive, rough, or clumsy by others.

Missing balance sense and the resultant problem of disequilibrium leads to significant motor delays. Postural security and a good sense of equilibrium depend upon the effective development and functioning of three different but interdependent sensory systems (an “Equilibrium Triad”), namely the vision sense, the balance sense, and the tactile/proprioceptive senses. In children with CHARGE all three of these sensory systems are likely to be missing or impaired, which largely explains the slow development of large motor skills and mobility, but also makes it remarkable that almost all children with CHARGE do eventually stand up and walk. Any input that helps to improve the functioning of any of the senses in this Equilibrium Triad can, therefore, be regarded as making a contribution to the development of independent standing and walking. For example, hippotherapy, or therapeutic horseback riding has been found to improve energy, walking, running, and jumping [McGibbon et al., 1998], and has been successfully used with children with CHARGE [Kruger, 2000].

There are strong links between the balance sense and vision [Glimcher, 1999] and problems with balance may affect the ability to maintain a stable visual field, to follow objects smoothly with the eyes as they move, and to differentiate whether it is the object or oneself that is moving [Gregory, 2001]. Some children may appear to “go blind” if their postural security is too challenged, but may show some well-developed visual (and other) skills once they are flat on their back or side on a stable surface. As they get older, children may use residual vision to help them to stay upright (think about the Equilibrium Triad), compensating for having no balance sense by using the strong visual impressions made by horizontal

and, especially, vertical lines in a room (e.g., corners, the edges of windows, doors, table tops, and wall-mounted pictures). They may have much less equilibrium outdoors where these strong visual markers are largely absent or beyond their range of vision. One result might be a reluctance to go outdoors, for example during recess at school, and another might be an inability to perform certain tasks out of doors that are carried out very well indoors. For children who are reading, the use of a typoscope (a letter-box shaped frame) can help by isolating one single line of text at a time. Similarly, the use of large print on a computer might be very helpful to a student, not because their visual acuity is poor but because they need help to isolate the line of text on which they should be fixating.

There may also be links between the balance sense and the ability to process sound and to develop spoken language [Colby Trott et al., 1993]. For children with CHARGE, this has implications in addition to other hearing difficulties, and a collaborative approach that brings together a Teacher of the Deaf, a Speech Therapist, and an Occupational Therapist trained in SI Therapy (or any combination of these) should be very helpful. Difficulties processing auditory input contribute to problems with memory and with learning some basic academic skills. We all need to move to some extent in order to listen, but children with balance problems may need to move even more to listen and to understand, so that telling them to “Stand still and listen” could be counter-productive.

Regular input from a Physical Therapist is very important for all children with CHARGE, but these therapists will need to be informed about the possibility of severe balance problems in this population and about the implications of this [Blake and Brown, 1993; Admiraal et al., 1998; Gregory, 2001].

In addition to visual difficulties that may result from a dysfunctional balance sense, specific ocular defects associated with CHARGE will also have a significant impact upon visual abilities. Colobomas of the retinas will cause some visual field loss, mostly in the upper visual field. As a result a child may at first appear to be completely blind or may just stare obsessively at bright lights. Then they may like to look at things “upside down” (in supine with the head tilted back and the object above the top of their head). Later, when upright and mobile, a child may have to tilt the head back in order to see in front of them—this posture might help with visual orientation during walking but it prevents children seeing where they are placing their feet, and it is very challenging for good sitting and standing posture and secure equilibrium. There may be extremely conflicting needs with regard to head position when walking—good equilibrium, the need for a clear view of where the feet are being placed, and photophobia may all compel the child to hold the head forward with the face down, yet the head needs to be held back with the face up in order to really see the environment. If retinal colobomas are located at the macula or the optic nerve then visual acuity will be affected. As a result of combined field loss and poor central vision children may not

look directly at objects or people, even though they are, in fact, looking. Retinal colobomas carry a high risk of retinal detachment, so high impact activities are not recommended. Extra care is also needed when imposing large rhythmic movements on children or doing gym activities. For older students, the risks of certain sports (trampoline, boxing, diving, wrestling, football, etc.) will need to be considered. Investigation of any dramatic change in a child's functioning should automatically include an ophthalmic examination [Pagon, 1999].

Visual acuity and visual fields should not be affected by colobomas of the iris, but this anomaly is likely to create problems coping with certain levels of light (photophobia), problems that can also sometimes be present even without iris colobomas. Indicators of photophobia may include screwing up the eyes or covering them with an arm or hand, holding the face down towards the floor all the time when outdoors in daylight, resistance to going outside in daylight, refusing to sit facing towards windows in the classroom, and refusing to face brightly illuminated computer screens. Provision of tinted spectacles or a sun visor or peaked cap can often ameliorate many of these problems. One apparent paradox is found when children who demonstrate photophobic behavior when they need visual information also at other times deliberately gaze at bright light when they only need visual stimulation (which could be an indicator of the need to get sensory systems reorganized due to tiredness, stress, or sensory overload). In other words, bright light can be great when it is just what the child needs, but it is a terrific nuisance to them at other times.

Facial palsy (cranial nerve VII) is another common anomaly with implications for vision. If the facial anomalies are marked, it is possible for one eye not to open very wide (ptosis), and/or the lid on the other eye never to close. With ptosis, the child will need to tilt the head back in order to see under the upper lid, possibly even using a finger to push the lid more open. If the eye does not close, artificial tears will need to be used to prevent drying out and scarring of the cornea [Pagon, 1999]. Facial palsy can also result in a very inexpressive face, which adds to existing difficulties with expressive communication, and often leads to incorrect or lowered expectations in others. A combination of bilateral facial palsy and macular coloboma (central vision loss) leads to a child with no facial expression who does not appear to make eye contact, which compounds other difficulties with clear expressive communication.

Ear anomalies and hearing difficulties may not be linked in CHARGE, but can be discussed together. Tentative diagnosis of CHARGE can occasionally be made just from the distinctive deformities of the external ear [Thelin et al., 1999]. Sometimes the shape of the external ear can be modified after birth using non-surgical procedures. More often surgery is used later in

childhood to facilitate hearing aid use, or for purely cosmetic reasons. Because of the floppy, deficient ear cartilage, surgery is not always successful. The ear canal can also be very narrow, so that temporary blockages are common. These anomalies cause problems with fitting ear molds and keeping hearing aids in place. This can be compounded by the child's preference for being supine, and ingenious solutions may have to be explored (e.g., Huggie Aids or sticky tape), or the use of bone conduction aids considered. Children who spend extended time on their backs are also at increased risk of oral and nasal secretions running into the ears, mucking up the hearing aids, causing blockages and infections in the ear canal. Noisy congested breathing is another common feature of CHARGE that has implications for the child's ability to perceive and respond to sounds in the environment.

It is common for the bones of the middle ear to be malformed [Dhooge et al., 1998], thus causing a significant degree of conductive hearing loss on top of that caused by fluid accumulation in the middle ear [Thelin et al., 1999]. Many children's long-term preference for being in the horizontal position also increases the chances of fluid build up in the middle ears. The resulting complex conductive hearing loss may require a high level of amplification. High levels of amplification increase difficulties with feedback because of poorly fitting ear molds and unhelpful postures.

In addition to the conductive hearing loss, most children with CHARGE have a sensorineural hearing loss due to malformations of the cochlea [Dhooge et al., 1998; Thelin et al., 1999]. Cochlear implants are now being carried out on some children with CHARGE with varying degrees of reported success [Weber et al., 1998; Stjernholm, 2003].

There are specific central nervous system anomalies associated with CHARGE, including anomalous auditory nerves (cranial nerve VIII), which connect the cochlea to the brainstem and the brainstem to the brain. Such abnormalities may cause Central Auditory Processing Disorder (CAPD). The meaning and implications of these anomalies are still being investigated [Thelin et al., 1999]. The most commonly seen manifestations of CAPD are difficulties perceiving one particular signal when there is competing noise, and problems with processing and understanding speech.

SI dysfunction appears to be inherent in CHARGE, and significant difficulties caused by impaired and poorly modulated sensory systems are very common. Many behaviors, some of them apparently contradictory, could indicate the need for SI assessment and treatment by a trained Occupational Therapist. These are listed in Table II.

Some typical techniques suggested by a therapist following an SI assessment include brushing protocols, rhythmic joint

TABLE II. Signs of Sensory Integration (SI) Difficulties in Children With CHARGE

- 
- Rejecting of textures in the mouth apart from pureed food, but mouthing of all kinds of non-food items, for example, stones, wood, cloth, soil
  - Absence of chewing and biting on solid foods, but excessive chewing and biting on non-food items, often with persistent teeth grinding
  - Rejecting certain tactile inputs as if they are painful, but apparent non-awareness of certain other tactile inputs (which for others might be painful!)
  - In the early years, extreme postural insecurity when placed in a sitting or standing position by an adult, or when moved unpredictably, but pleasurable responses to strong rhythmic movement experiences (e.g., rocking, bouncing, swinging)
  - Severe problems with regulating arousal levels, often described as periods of frantic over-activity and over-excitement and stress, but also sudden periods of apparent "burn-out"
  - Abnormally high pain thresholds
  - Inconsistent or inappropriate use of pressure when touching or grasping with the hands, often described as the child being very "rough" or "clumsy" or "aggressive," and generally poorly graded movements
  - Very delayed awareness of bowel and bladder movements
  - Disturbed and inconsistent sleep patterns
  - Behaviors that seek and provide very strong sensory inputs like self-biting or scratching, skin picking, spinning, rocking, bouncing, shoulder shrugging, leg swinging, hyperventilating, hand flapping, self-slapping, as a way of getting the body reorganized
-

compression, deep tissue massage, vibro-tactile input, a range of large movement activities, and the use of weighted clothing. A variety of outcomes might be anticipated from implementing these techniques, including improving the child's ability to attend to sensory information in the environment, improving muscle tone, improving awareness and tolerance of touch, improving attention span and decreasing distractibility, reducing the need for self-injurious behavior, improving sleep patterns, and generally increasing the child's ability to remain both alert and calm in stressful situations [Wilbarger and Wilbarger, 1991; Stock Kranowitz, 1998; Deuce, 2002; Larrington, 2002].

This kind of SI perspective might be needed, regularly or periodically, throughout the individual's life and should never be automatically regarded as a one-time "fix." Experience strongly suggests that every person with CHARGE would benefit significantly from having a regular SI program under the supervision of a suitably qualified Occupational Therapist. When requesting an SI evaluation it is important to list the precise behaviors that have led to a suspicion of sensory integrative difficulties so that the therapist will be helped in advance to know what the key issues might be [Maynard, 2001].

Many children with CHARGE need extended time to process information, and often develop techniques that they use to establish a firm physical, emotional, perceptual, and cognitive "base" each time before they can respond. Successful teaching frequently depends on allowing for this need and spending some time alerting the child to the fact that you are there, who you are, what you are going to be doing together, how and where it will be done, and so on. The child may need considerable time, and assistance, in establishing a secure and stable physical base as a first priority in every communicative interaction.

Any and all modes of communication may be appropriate for children with CHARGE. Each child must be considered as an individual, but exposure to a variety of communication modes (especially including those with a concrete component such as symbolic object systems, pictures, or picture symbols) is usually helpful, so that they can eventually make a choice of their preferred modes, which might be different for reception and expression. As an example, some children learn to understand spoken English (often with amplification), but express themselves predominantly with sign language—this should not be considered bizarre or illogical but merely a reflection of the way that each of the CHARGE anomalies in Table III may pose obstacles to the production of clearly articulated speech.

In spite of this daunting list, speech is often the eventual preferred mode of expression for a surprisingly large number of children, though they may also use signs or picture symbols expressively as additional support because of poor speech articulation. Many of the CHARGE anomalies, quite apart

from vision and hearing impairment, also carry implications for the comprehension and expressive use of sign language. Receiving signs tactually, and expressing with signs, may be compromised by under-functioning tactile and proprioceptive senses, low muscle tone, severe balance problems, and dyspraxia, which is thought to be a problem for many children [Maynard, 2001; Nicholas, 2005]. Central Auditory Processing Disorder may compound difficulties with perceiving and understanding spoken language.

Many people with CHARGE, including those who seem to have good levels of language, demonstrate difficulties with vocabulary recall, initiating communicative exchanges, and with clearly articulated expression, in the abstract forms of spoken and/or sign language. Problems with initiation may also result from specific brain anomalies [Nicholas, 2005]. Provision of a communication mode with a concrete component (e.g., objects, symbols, pictures, written words), possibly in the form of a Communication Book, can be of immense help in aiding recall, in encouraging initiations, in clarifying meaning, and in generally fostering a more confident, animated, and fluent communicative style.

The following anecdotes of children with CHARGE syndrome are examples of behaviors that were considered to be "challenging" by family or school that were removed, or moved into the "non-challenging" category, by taking a multi-sensory view, based upon our knowledge of all the sensory difficulties associated with CHARGE. In many of these examples, there was a clear need for SI assessment and programming along with the other strategies mentioned:

- A young child who was said to be on his back self-stimulating "all the time" was actually practicing and developing his mobility and orientation skills, and using vision and touch to explore objects, very creatively. While doing this he needed to get onto his back on the floor every 10–20 min to reorganize his sensory system with brief episodes of limb shaking and hyper-ventilating.
- A kindergartner was often self-abusive when he got distracted and over-aroused by incidental touch and air movement caused by people repeatedly walking behind his chair. Once his chair was placed with its back securely against a wall he was less self-abusive and more amenable to social interaction.
- People were concerned when a young boy began to insist on the unusual idea of wearing band-aids wound tightly around the tips of all his fingers and thumbs every day. He was expressing his need for more and stronger pressure and touch inputs as a part of his sensory diet, inputs that helped with postural control and mobility as well as with fine hand and finger skills.
- Every morning in a pre-school program a student refused to sit on the floor with her class to watch the teacher sign a story. When an appropriate chair was provided the student sat and attended with great interest and a growing level of participation.
- Many children who were unable to sit on a regular chair and attend to an activity for very long showed an extended attention span and better visual, fine motor, and cognitive functioning once given chairs with footrests and armrests. One child rarely used the armrests for his arms but instead sat with his legs spread wide and his outer thighs pressed hard against the sides of the seat, giving him the requisite equilibrium to function effectively in the upright seated position.
- A girl was described as very disruptive during sessions that required the class to sit still and participate in a signed conversation with the teacher for up to 30 min. When the teacher used a strategy of asking the student to move periodically to carry out small chores during these sessions

TABLE III. Obstacles to Clear Articulation of Speech in Children With CHARGE

- 
- Hearing impairment
  - Vision impairment
  - Facial palsy
  - Low muscle tone
  - Poor tactile sense
  - Oro-facial clefting
  - Enlarged tongue
  - Poor tongue movement
  - Small lower jaw
  - Larynx and pharynx anomalies
  - Breathing difficulties
  - Swallowing difficulties
  - Dental abnormalities
  - Extremely delayed/immature eating skills
-

(to fetch a pen, open a door, bring a book, take a paper to the school office) the disruptive behaviors largely ceased.

- A teenager enjoyed, and was quite good at, soccer in the school gym, but was unwilling or unable to play it outdoors due to the absence of strong vertical visual markers to aid equilibrium. The ability to participate in such complex physical activities outdoors did not develop until significant adaptations were introduced.
- A different teenager was unwilling to go outside during school recess because of problems with glare and photophobia that impacted mobility and orientation, as well as participation in signed conversations. This difficulty was eventually solved by the provision of tinted glasses and a sun visor.
- During Orientation and Mobility sessions a teenager was refusing to stand still to receive spoken/signed instructions, but the problem was solved when the student was allowed to stabilize himself by leaning against a pole or a tree or a wall, or by placing one hand on the instructor's shoulder during these conversations.

There has been a long-standing debate about "CHARGE behavior" amongst families and professionals, and now a more decisive focus on behavior is emerging in several countries. I would want to remind people of the immense difficulties that children with CHARGE face in almost everything that they do, and, as a consequence, of the very high levels of stress with which they must live for much or even all of their lives. Time spent trying to reduce stress levels, and trying to give the children acceptable strategies for doing this for themselves, must be one of the most precious gifts we can offer them, and one of the biggest favors we can do ourselves as family members, educators, and therapists.

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# Welcoming a Child with CHARGE Syndrome into Your Classroom

## A Behavioral Perspective

By Laurie S. Denno, MA, BCBA  
Behavior Analyst

Children with CHARGE syndrome are first and foremost children. While their genetic condition is rare and their sensory impairments can be daunting, for the most part their behavior challenges are similar to other students. They may fuss or have a tantrum if they do not get what they want; they may throw materials or cry if their work is too hard; and they may engage in any number of disruptive or undesirable behaviors to get attention from adults and other students. These behavior issues generally respond well to an appropriate treatment based on a thorough functional analysis of behavior. A behavior analyst should be sought to help sort through these challenges. A behavior analyst will observe the student and talk with the educational team and family. The behavior analyst will then offer recommendations for treatment, design a behavior plan, train staff and family members to implement the plan and provide on-going monitoring, follow up and treatment adjustments based on outcomes. To be most effective, behavior analytic treatment requires a regular on-going commitment. If you do not have access to a behavior analyst, you can obtain the name of a behavior analyst by going to [www.BACB.com](http://www.BACB.com) and searching for a Board Certified Behavior Analyst by your city or zip code. You also can call organizations that serve developmentally disabled children and adults such as the ARC, Easter Seals Foundation, children's hospital or university hospital developmental disabilities unit and special education or psychology departments at a nearby university.

Students with CHARGE syndrome may display a number of unusual behaviors that appear somewhat different from the behaviors noted above. Some may be significant enough to require extra treatment, such as psychiatric and/or medical intervention. These behaviors can include:

- a high level of anxiety or nervousness
- repetitive questions about the same topic
- inflexible behavior
- upsets over changes or disruptions in schedules or routines
- repetitive non-functional behaviors (OCD-like)
- aggression or self-injurious behavior as a result of redirection from repetitive non-functional behaviors
- withdrawal
- autistic-like behaviors
- sleep problems: both falling asleep and staying asleep

The best way to address these behaviors is to work together with the family to design a consistent response to the behaviors. Communication about schedule changes,

repetition of the facts, rules and reinforcement can help with anxiety over routines and changes in routines. Time to relax or regroup (this is not time out – see article by David Brown) can alleviate many behavioral difficulties. The best approach appears to be to “work around” many of the behaviors rather than trying to suppress them. Generally speaking, negative feedback, reprimands, physical redirection and punishment have not been found to be effective in treating these behaviors in these children.

## Checklist for Behaviors in CHARGE Syndrome

Martha M. Majors, M.Ed., Teacher of the Deafblind

Pam Ryan, M.A., CAGS, Licensed Educational Psychologist

All behaviors happen for a reason. Children with CHARGE syndrome have many complex factors that influence behavioral responses. Many sudden, unexpected changes in behavior are due to medical issues. The following is a simple checklist that can help staff working with these children to increase their awareness of some of the reasons why behaviors occur.

- Has the child had enough sleep
- Has the child had adequate nutrition
- Has the child been seen by health professionals for sinus, ear infections, constipation
- Has the child's schedule been designed to prevent fatigue
  - Has the child had several active classes in consecutive session and simply cannot pay attention for that long without a break
  - Does the child have both vision and hearing challenges and therefore has to work very hard to attend to communication and curriculum content, with the outcome of poor attention, outbursts, and /or the child simply shuts down
- Are the child's hearing/vision/sensory needs being met so she/he can function in the classroom
  - is text accessible
  - is there ambient noise
  - bothersome sun or outside noise
  - is his/her desk in the path of classroom traffic that might be distracting
  - are hearing aid batteries/FM unit working
  - are glasses in good repair and clean
- If the child is relying on sign language, is this available in all places the child goes to during the day
- Is there a back up communication system for times of stress such as pictures, needs board, notebook with short printed comments (take a deep breath, ask for help, ask for quiet time, etc.) Are these available to the child at all times
- Is there a flexible schedule

Behavior responses of the child to any or all of the above might include:

Increase in anxiety

Increase in OCD types of behaviors and thoughts

Increase in asking questions about what is next, what is cancelled, etc.

Increase in overall arousal levels

Increase in tantrum, shut down, or general agitation

Decrease in the ability to clearly communicate and to be an effective self-advocate

Self-stimulatory behaviors: picking at nails, skin, hair; body movements

## **Physical Characteristics of Children with CHARGE Syndrome**

Maryann M. Girardi, PT, DPT, ATP

Children with CHARGE syndrome have physical characteristics which affect their gross motor performance. These characteristics interact with each other to affect the child's balance and ability to interact with their environment. The following is a brief discussion of these characteristics, their interactions and some accommodations for them:

### **Low muscle tone**

Muscle tone is defined as the "resistance to passive stretch." What this means is, when the muscle is moved, does it resist the movement (high tone), does it go along with the movement (normal tone), or move quickly without tension (low tone)? Muscle tone is involuntary and should not be confused with strength. Low tone muscles are slower to contract and unable to maintain a contraction for as long as normal tone muscles. This means children with CHARGE syndrome may not be able to sit for as long as their peers. They need position changes approximately every 30 minutes. These changes may include standing and working at a higher surface (so they do not have to bend over), lying on their stomachs on the floor (for a more stable position) with work in front of them, and sitting in a more supportive chair such as a bean bag or recliner to decrease the amount of physical effort required to sit up.

### **Visual Impairments**

Decreased visual field (blind spots, loss of upper visual field with retinal colobomas) and decreased acuity are among the visual impairments that affect motor performance. Visual information is used to guide muscle performance and movement as well as being a component of balance. When there is decreased visual information, the quality and/or ability to move will be affected. Consultation with a vision specialist can help in understanding the visual impairments and in planning for specific accommodations. Accommodations may include adequate lighting, increased contrast for edges/ borders of objects, and special paper with large lines.

### **Skeletal Anomalies**

Spinal anomalies are sometime present, including congenital cervical spine fusions, kyphosis and scoliosis. Many have torticollis and decreased use of the shoulders. These will affect the ability to:

- rotate the head, resulting in having to turn the trunk as a whole to see from side to side
- raise the head to look up (to accommodate for loss of upper visual field)
- maintain an upright trunk with the head in midline
- raise arms – often they cannot raise their arms above the shoulders

Materials should be presented within the physical limits of the child's posture. Adaptations to the classroom seating will be necessary to allow full access to materials. Consultation with physical and occupational therapists for modifications specific to the student would be helpful.

## **Vestibular System Impairments**

The nearly ubiquitous vestibular system impairments in children with CHARGE inhibit the ability to detect body motion and position in space. This is important for maintaining an upright posture and is an integral part of the balance system.

## **Balance**

The body's ability to balance is a complex mechanism, normally using input from vision, muscles, joints and the vestibular system. All of this information must be processed by the brain and translated to motor output (movement). Children with CHARGE syndrome have impairments in all of these areas. They have decreased visual and vestibular information. Because of the low muscle tone and skeletal anomalies, the information from the muscle and joint receptors is also decreased. The ability to process this information may also be decreased. The resultant motor output is also affected by the decreased muscle tone and visual impairments. The motor response time may be increased. The stability of the joints is decreased and difficult to maintain. The visual information is not present to direct movement. Sometimes the kids just fall down or need to lie on the floor for a while! Ability to move about may be different indoors (with walls to serve as visual cues) and outdoors, due to differences in visual cues (walls, doorways) and surface changes (floor vs. grass).

Accommodations can be made to increase the sensory information available which can in turn increase the ability to balance. These include increased lighting and the use of contrasting colors to define the edges of stairs, curbs, or other surface changes to increase the visual information. Provide safe environments with increased physical assistance to ensure success while performing activities that require balance, such as walking on a balance beam (which has been modified to increase visual information), roller skating, scooters. Help enable success and repetition with decreasing assistance, effective balance reactions can be "learned." Consultation with physical therapists to provide student specific accommodations can be helpful.

## **CHARGE Syndrome Information: Occupational Therapy (OT)**

Anna Shinkfield, OTR/L

When working with a student with CHARGE syndrome, many of the typical occupational therapy treatments/strategies need to be altered or modified.

For example, it is beneficial for the student if sessions are set up with:

- A set routine from session to session
- A visual schedule [pictures or symbols] in order for the child to decrease anxiety and to anticipate what will come next
- Provide a “finish bucket” for the child to place the completed activities in
- Provide minimal visual and auditory distractions
- Set-up sensory breaks throughout the session

Every student with CHARGE is unique and requires a full team evaluation to determine what adaptations and modifications are likely to be most successful. Ideally, the full team would include parents, classroom teachers and aids, speech language pathologists, physical therapists, and other educational specialists. In addition, information provided by numerous medical specialists (e.g. ophthalmology, audiology, neurology, cardiology) should be available and taken into account. If possible, a behavior specialist should be part of the team. A behavioral specialist may offer suggestions for ways to set up the environment, as well as how to address any negative behaviors.

Some of the conditions present in CHARGE which might benefit from OT attention.

- bone and muscle malformations of the hand
- increased tightness and shortening of the muscles in the shoulder area
- low tone resulting in loose joints
  - difficulty holding a pencil or pen
- upper body hypotonia
  - tiring more quickly when compared to their peers
- decreased shoulder mobility (many cannot lift their arms above shoulder level)

These abnormalities may inhibit the child’s ability to complete many activities, including activities of daily living (ADL’s), finger spelling and signing, handwriting tasks, and other fine motor classroom activities. It is important to consider what is being asked of the child, and if they are physically able to perform that task. Knowing the limitations of each child is critical in planning activities for that child.

Another area that may need to be tailored differently for a child with CHARGE syndrome is the basic structure of their day. In order for them to be involved in their schedule, and to anticipate and be prepared for what comes next, try to keep to a set routine during the day and for therapy sessions. Collaboration with the SLP and other therapists is encouraged to incorporate the most successful means of total communication for setting up the schedule.

## Speech and Language Therapy in CHARGE Syndrome

Lisa Anne Guerra, MS, CCC-SLP

Students with CHARGE syndrome benefit from speech and language therapy to expand their receptive/expressive language skills, social/pragmatic language skills and in some cases speech production skills across settings. Assessing and treating a student with CHARGE syndrome differs from typical evaluation and treatment protocols used by speech and language pathologists. It needs to be a team approach, ideally involving the classroom teacher, parents, teacher of the visually impaired, audiologist, occupational therapist, and physical therapist. The SLP needs to integrate the visual, auditory, sensory, behavior, and educational information for each individual student based on the assessments from the respective disciplines.

A ***Total Communication*** approach works best for these students. *Total Communication* means incorporating spoken language, vocalizations, objects, tangible symbols, body language, gestures, sign language, tactile sign language, photographs, line drawings (e.g., Mayer-Johnson picture symbols), and written text/Braille depending on the individual student's profile. The total communication approach should be used to convey information to the student and for the student to use *to communicate*. It should be stressed to parents that learning sign (often the easiest for a child to learn first) does not mean a child will not learn to speak – use every available means of communication!

The speech and language pathologist needs to look at the student with CHARGE syndrome as a whole person, taking into account structural, anatomical, and physiological characteristics of the syndrome, as well as the positioning, sensory needs and behaviors of the child. Many times a student is unable to attend and/or to carry out a task because of the features of CHARGE. This can be misconstrued as non-compliance or lack of understanding when it is neither. For example, when presented with a novel activity, the student may want to explore the activity on their terms and may not be able to follow the task requirements until the activity becomes more familiar.

The student with CHARGE benefits from ***established routines***, where predictability helps decrease anxiety and thereby ensures better availability for learning. Working closely with the team will allow the speech and language pathologist to establish a positive rapport with the student and allow for maximum communication growth on the student's part.

## **CHARGE Syndrome: Technical Assistance (TA) for Children with Combined Hearing and Vision Loss (Deafblindness)**

Betsy McGinnity, M.Ed.

Most children with CHARGE syndrome have varying combinations of hearing loss and vision loss. This is referred to as deafblindness even when there is some residual vision and/or hearing. Children with deafblindness require distinct educational practices to assure that they reach their potential. That is, approaches used with children who only have hearing loss or only have vision loss are not sufficient. Even children with some usable vision or hearing will receive distorted or incomplete information through these senses. To benefit from their education, children with CHARGE require significant support and curricular modifications.

Most children who are deafblind are educated in their local school districts. Because deafblindness is such a low incidence disability, the local district may never have served a student who is deafblind and will lack the local expertise to do so. ***Technical assistance from both state and national Deafblind Projects are the primary way for local districts to get the training and support needed to effectively educate these children.*** Technical assistance is a process of linking and applying state-of-the-art knowledge & practice to the solution of a [developmental] problem. Technical assistance is designed to help clients develop or improve systems, organizations, programs and or products (NECTAC, 1989.)

The state and national projects focused on deafblindness support efforts to train both beginning and veteran teachers about the unique needs of students who are deafblind, about appropriate assessment and effective teaching strategies.

### **State Deafblind Projects**

- serve children and their families in each state. For help finding the project in your state contact DB-LINK (800) 438-9376) [www.dblink.org](http://www.dblink.org) [People and Programs tab]
- offer training, support and consultation to teachers, other educational team members and families
- work with State and Local Education Authorities to help them provide appropriate services for children who are deafblind
- often employ a Family Specialist to provide support and networking opportunities for families

### **National Consortium on Deaf-Blindness (NCDB) <http://nationaldb.org/>**

- works, nationally, to identify the needs of children and youth who are deafblind
- delivers an array of technical assistance to meet the needs of families, service providers and agencies

- strengthens the capacities of service providers and agencies serving children and young adults who are deafblind and their families
- increases the ability of families to effectively participate in their child's education
- links research-based practices to state and local service providers and families

**DB-LINK at NCDB** <http://nationaldb.org/ISDefault.php>

- Facilitates the sharing of information and practices between agencies, organizations and individuals
- Provides resources and personalized service to anyone needing information about or for children who are deafblind through a toll free number (800) 438-9376 and on the web
- Helps parents, teachers, and others by providing them with information to foster the skills, strategies and confidence necessary to nurture and empower children who are deafblind
- Identifies, coordinates, and disseminates, at no cost, information related to children and youth who are deafblind from birth through young adulthood

## **CHARGE Syndrome Information: Vision**

Donna Bent

Certified Orientation and Mobility Specialist

Certified Teacher of the Visually Impaired

Children with CHARGE often have vision impairments due to colobomas. Most children with colobomas have a visual field loss, typically in the superior (upper) field. There may be blind spots and acuity problems in addition to the visual field loss. Iris colobomas can result in extra sensitivity to bright lights, even indoors. In many cases, one eye is more severely affected than the other, causing monocular vision and lack of depth perception.

Understanding (as much as possible) the extent of the child's "visual bubble" (how far out and how well can the child see) is critical for educational planning. A child who can pick up a paper clip on the table may not be able recognize the teacher at the front of the room or even facial expressions at relatively close range. A child with CHARGE probably also has significant hearing impairment and vestibular (balance) abnormalities, both of which also impair the amount and quality of information received from the environment. Ideally, a DeafBlind specialist can be consulted to help understand the complications associated with dual sensory impairment [see Technical Assistance].

A Certified Teacher of the Visually Impaired (CTVI) can help provide ongoing functional vision assessments and make recommendations for improving visual skills. Accommodations should be made for safety as well as for the visual enhancement of the environment. Even if the child does not appear to be severely visually impaired, learning can be challenging due to a combination of factors specific to CHARGE as well as the additional sensory losses of hearing impairment and balance issues.

With a field loss or monocular vision, it is difficult to follow a line smoothly or a series of lines systematically. Reading or any type of precise near reading work is both challenging and fatiguing. Large print, bold lines, underlining, or a line marker can be helpful.

A visual field loss also requires a child to scan systematically in order to receive the full visual picture. This can make following a signed conversation and receiving all the information quite exhausting. In a traditional educational setting, visual information may come from signing (distance), handouts (close up) and computer projections or a chalkboard (distance). Children have difficulty processing all of this information together in a coherent manner. It is also exhausting for the child to do this for extended periods of time. Teachers of the Visually Impaired can help with compensatory strategies for accommodation and strengthen visual skills. The CTVI can also assist the classroom teacher in adapting materials and the environment to make learning more successful.

Children with CHARGE typically have difficulty moving through space and learning through the visual sense. A Certified Orientation and Mobility Specialist (COMS) can help people with vision impairments to learn to move through space more safely and

efficiently. Almost all individuals with CHARGE have difficulty maintaining balance. There are specific Orientation and Mobility techniques that help compensate for the poor balance. Visual field deficits affect safety while moving as obstacles or drop offs may be missed. Many children with CHARGE have difficulty transitioning in different environments. Orientation and Mobility training provides structure and specific training that make the transitions more efficient and safe.

## **CHARGE Syndrome Information: Oral-Motor and Feeding Issues**

Anna Shinkfield, OTR/L

Lisa Anne Guerra, MS, CCC-SLP

Most students with CHARGE syndrome exhibit oral-motor and/feeding issues. These students benefit from therapy with speech and language pathologists (SLP), and occupational therapists (OT) working together. The oral-motor/feeding therapy team may also involve parents, pediatrician, gastroenterologist, nurse, behavior specialist, physical therapist, psychiatrist/psychologist, as well as classroom staff. Consistency of routine and communication among everyone dealing with the child is important.

Due to the complex oral motor/feeding profile of these students, as well as the strong emotional component around mealtime, it is imperative to assess and treat the child as a whole. Because feeding is a multisystem issue, assessment needs to be done with a team of the above-mentioned professionals. Once the medical team has determined that the student can eat orally, an assessment will look at the following: nutritional needs (calories), structure and function of the oral cavity, sensory awareness in the oral cavity, positioning, and ability to tolerate various textures/consistencies/temperatures. A modified barium swallow study (a procedure in which the process of swallowing and the movement of food from the mouth through the esophagus is viewed in order to determine how well the airway is protected during swallowing) is likely to be part of the initial evaluation. The SLP and OT can work together to develop a treatment program, taking all of the assessment information into account. Ongoing discussion with the complete team is warranted in order to measure progress.

### *FROM THE MANAGEMENT MANUAL FOR PARENTS:*

CHARGE syndrome affects the cranial nerves, especially those involved with oral-motor skills and swallowing. Many children remain primarily g-tube fed for years. Those who eat orally often continue to have difficulty with certain textures or consistencies of food. Choking may remain a potential hazard forever. Secondary factors leading to oral-motor/feeding issues include gastroesophageal reflux (GER), cardiac and respiratory complications.

Reflux can lead to aspiration pneumonia and increase the frequency of ear and sinus infections. Pain associated with infections can be the underlying cause of abrupt changes in behavior in children with CHARGE. When sudden changes in behavior (including apparently aggressive or self-injurious behaviors) are seen, medical causes of pain should be ruled out before instituting behavior modification programs.

## **Educational Needs of Children with CHARGE Syndrome**

Martha M. Majors, M.Ed.

Sharon Stelzer, M.Ed.

Educating a child with CHARGE is very complex. When developing an educational program many components should be considered. These include the following topics:

### **COMMUNICATION**

Most children with CHARGE have both vision loss and hearing loss (dual sensory impairment: deafblindness). Most benefit from a **total communication** approach. Total communication means incorporating anything and everything: gestures, simple signs, print/Braille, facial expression, symbols, and PECS (Picture Exchange Communication System) in addition to speech and sign language. The modes for each child are highly individualized and the entire team (including the family) needs to participate in developing and consistently instituting the plans.

### **SENSORY LOSSES AND THE IMPACT OF VISION AND HEARING LOSS**

It is important for the team to take careful notice of the functional vision of the child as well as the hearing loss, the impact of hearing loss on communication and the total effects on day-to-day functioning. Determine the communication bubble! [The communication bubble includes how far out and how well a child sees and how far out and how well a child hears. It also needs to take into account things like lighting, ambient noise and other factors.] Many types of accommodations will likely be necessary in the educational setting and these should be written into the child's IEP. Consultation with vision and hearing specialists can be helpful in determining appropriate accommodations.

### **CURRICULUM**

Curriculum should be challenging, and both age and content appropriate. Children with CHARGE are very capable. Due to communication challenges and delays in using expressive language, their overall cognitive ability is often, if not usually, underestimated. In fact, many children with CHARGE are very clever, have an impressive capacity to learn information, to retain ideas and to do well in academic subjects. They have specific learning needs, can benefit from specific teaching strategies and must be challenged through access to a highly motivating and flexible curriculum. They benefit from a curriculum that incorporates a variety of skills across a variety of settings. As goals are achieved, other skills can be added to enhance knowledge and growth.

### **ENVIRONMENTS**

Children with CHARGE often require frequent sensory "breaks." Some types of break examples of breaks might include:

- bean bag chair
- soft cushion on the floor
- magazines and books in a comfortable space

- quieter room with low lighting; small space with three walls and comfortable seating
- swings of different types
- weighted blanket or vest

There should be adequate space within classrooms or nearby to ensure access for the child who needs time to relax, to do something that calms and allows time to be quiet (see article by David Brown). Fatigue can quickly overcome a child. The ability to take a quick “break” is critical for the child to be able to maintain a calm demeanor. A flexible schedule that allows for “breaks” is important; staff should be aware of “signals of behavior” and how to respond to the child; anticipation of the need for a break is a key to minimizing the behavioral outbursts and creating a successful educational environment for a child with CHARGE!

Physical environments can be arranged to motivate and intrigue the child with CHARGE. By having a flexible environment that can change and grow, optimal learning can take place. For example:

- Having a space that reflects the likes of the student, such as hanging pictures of their favorite people including characters can make the work or relax space inviting.
- Having some environments or spaces created with intriguing materials that peek the interest of the student can make learning fun.
- Rotate materials to keep the child interested and motivated.
- Using creative imaginative ideas can change an ordinary space into anywhere in the world. For example, the science area can become a space ship to our solar system.

## **TEACHING STRATEGIES FOR CHILDREN WITH CHARGE SYNDROME**

As far as possible, these ideas should be integrated into all educational settings and across all environments:

- Strategies for structuring activities
  - Organizational skills – work in an organized manner (left to right, top to bottom), using checklists
  - Negotiation – allow the child to feel as if he/she is in control. Give a variety of choices as well as use first, then, strategies.
  - Sharing – foster peer-to-peer interactions. Encourage turn taking as a precursor for conversational communication.
  - Motivation – select activities that are interesting to the student. Use materials that are interesting and rewarding.
  - People preferences – give choices of WHO should do tasks/lessons with the student whenever possible.

- Partial vs. Full Participation – have the child do as much of a task or activity as possible to feel successful. Avoid setting up for failure by having an activity that has too many steps or is too long.
- Functional Activity – in all activities, ask yourself “will this skill or activity be useful in the future?”
- Modeling – engage the child by modeling the steps in an activity
- Choice making – allow for choices across settings. Let the child feel in control. Choices may be limited or guided, but must be offered.
- Age appropriate content/curriculum – all content and curriculum should be based on age and cognitive abilities. Don’t underestimate the cognitive abilities of these children! Both factors should be considered when planning.
- Active vs. Passive learning – alternating activities between quiet (sitting-desk work) and movement oriented (delivery jobs, getting things, physical education class, recess, etc.)
- Clear expectations/Limit setting – set clear, firm limits that remain consistent. Give the child time and opportunities to learn these expectations.
- Follow Student’s Lead – allow students to be leaders, watch and see what interests them. Incorporate these ideas into lessons.
- Natural routines environment – teach in environments in which activities naturally occur.
- Variety of exciting content – although routines and structure are important, keeping the content dynamic is critical for the student with CHARGE!
- Task analysis – break tasks into smaller steps that allow the child to feel successful.
- Sensory techniques
  - Awareness of hands/touch – allowing others to touch for signaling, (tapping) tactile signing, etc.
  - Signals: Verbal/Auditory/Visual – gesturing to gain attention, using voice or sound cue to gain attention
  - Prompt levels hand-under-hand – use an adults’ hand as guides under the child’s as a less invasive technique.
  - Sensory breaks – allow pause time during activities and in between activities.
- Timing
  - Beginning-middle-end – make sure all activities have a clear beginning, middle, and end that are at the student’s level of understanding.

- Structure and routine – having predictable schedules and routines play to the strength of the child with CHARGE.
- Pause time for response – WAIT, WAIT, WAIT, allow the child with CHARGE to respond in a manner they are most comfortable.
- Curriculum
  - Child centered curriculum – the child should drive the curriculum, the curriculum should not drive the child.
  - Expanding environments – some children with CHARGE need to start activities in “smaller” environments (less physical space or less cluttered). As they mature and grow environments can become bigger (physically) and more challenging.

### **What Every Student with CHARGE Needs to Know:**

- Social Skills (how to be an effective communicator)
  - How to be a part of a group – as member that is accepted by the group
  - How to Negotiate – how to get their point across and know when to surrender
  - How to take turns & share – to take the appropriate amount of turns in a conversations—BE a listener as well as a speaker signer.
  - How to help out – be part of a family by doing chores. In school have school jobs.
- Cognitive Skills
  - How to be organized – work, think & execute in an organized manner
  - How to anticipate activities (calendar system) – know exactly when the order things will happen
  - How to cope with behaviors – know when & what to do when on “sensory” overload or stressed.
  - How to make choices – know that here are choices & the choices may be from a “set” group. Know that all choices are not possible at all times.

**\*\*See teaching strategies\*\*\***

## **CHARGE Syndrome: Resources for Teachers**

Betsy McGinnity, M.Ed.

### **Education**

Source: Minnesota DeafBlind Technical Assistance Project

<http://www.dbproject.mn.org/education.html>

### **Teaching Strategies and Content Modifications for the Child with Deaf-Blindness**

Source: Texas School for the Blind and Visually Impaired

<http://www.tsbvi.edu/Outreach/seehear/archive/strategies.html>

### **Educational Practices**

Source: DB-LINK at the National Consortium on Deaf-Blindness

<http://nationaldb.org/ISSelectedTopics.php?topicCatID=9>

### **Environmental Considerations**

Source: DB-LINK at the National Consortium on Deaf-Blindness

<http://nationaldb.org/ISSelectedTopics.php?topicCatID=62>

### **Issues in Deafblindness: When the IEP Is Aligned with the General Curriculum**

Source: Texas School for the Blind and Visually Impaired

<http://www.tsbvi.edu/Outreach/seehear/spring06/issues.htm>

### **Team Structure for a Deaf-Blind Student**

Source: Deaf-Blind Perspectives

<http://www.tr.wou.edu/tr/dbp/pdf/sept00.pdf>

### **Literacy**

Source: DB-LINK at the National Consortium on Deaf-Blindness

<http://nationaldb.org/ISSelectedTopics.php?topicCatID=32>

### **IEP Quality Indicators for Students with Deafblindness**

Source: Texas School for the Blind and Visually Impaired

<http://www.tsbvi.edu/Outreach/deafblind/indicators.htm>

### **Considerations in IEP Development for Children Who are Deafblind**

Source: Pennsylvania Deafblind Project

<http://cdd.unm.edu/deafblind/forms/facts/IEPDevelopment.pdf>

### **Documenting Modifications in the IEP for the Student with Deaf-Blindness**

Source: Texas School for the Blind and Visually Impaired

<http://www.tsbvi.edu/Outreach/seehear/archive/modifications.html>