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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 fisted, 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 lead 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.

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

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

(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 on 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.

REFERENCES

Abadie V, Wiener-Vacher S, Morisseau-Durand MP, Poree C, Amiel J, Amanou L, Peigne C, Lyonnet S, Manach Y. 2000. Vestibular anomalies in CHARGE syndrome: Investigations on and consequences for postural development. Eur J Pediatr 159:569–574.

Admiraal RJ, Joosten FB, Huygen PL. 1998. Temporal bone CT findings in the CHARGE association. Int J Pediatr Otorhinolaryngol 45:151–162.

Blake KD, Brown D. 1993. CHARGE Association looking at the future–The voice of a family support group. Child Care Health Dev 19:395–409.

Colby Trott M, Laurel MK, Windeck SL. 1993. Sense abilities: Understanding sensory integration. Tucson, AZ: Therapy Skill Builders.

Davenport SLH. 1999. Overview of CHARGE syndrome for physicians. In: Hefner M, Davenport SLH, editors. CHARGE syndrome: A management manual for parents. Columbia, MO: CHARGE Syndrome Foundation.

Deuce G. 2002. Sensory integration dysfunction in deafblind children. DbI Rev 30:8–10.

Dhooge I, Lemmerling M, Lagache M, Standaert L, Govaert P, Mortier G. 1998. Otological manifestations of CHARGE association. Ann Otol Rhinol Laryngol 107:935–941.

Glimcher PW. 1999. Eye movements. In: Zigmond MJ, Bloom FE, Landis SC, Roberts JL, Squire LR, editors. Fundamental Neuroscience. San Diego: Academic Press. pp 993–1010.

Gregory BB. 2001. Physical therapy and occupational therapy in CHARGE syndrome. In: Hefner M, Davenport SLH, editors. CHARGE syndrome: A management manual for parents. Columbia, MO: CHARGE Syndrome Foundation.

Hartshorne TS, Cypher AD. 2004. Challenging behavior in CHARGE syndrome. Ment Health Aspec Dev Disabil 7(2):41–52.

Kruger C. 2000. Personal account. CHARGE accounts, Vol. 10 No. 2, Summer 2000, 3.

Larrington GG. 2002. Sensory integration. In: Alsop L, editor. Understanding deafblindness: Issues, perspectives, and strategies. Logan, UT: SKI-HI Institute.

Maynard S. 2001. The impact of sensory integration dysfunction in CHARGE. In: Hefner M, Davenport SLH, editors. CHARGE syndrome: A management manual for parents. Columbia, MO: CHARGE Syndrome Foundation.

McGibbon NH, Andrade CK, Widener G, Cintas HL. 1998. Effect of an equine-movement therapy program on gait, energy expenditure, and motor function in children with spastic cerebral palsy: A pilot study. Dev Med Child Neurol 40:754–762.

Moss K. 1993. Looking at Self-Stimulation in the Pursuit of Leisure or I'm Okay, You Have a Mannerism. P.S. NEWS!!! 5(3) Published by TSBVI Deafblind Outreach Texas School for the Blind and Visually Impaired. http://www.tsbvi.edu/Outreach/seehear/archive/mannerism.html

Murofushi T, Ouvrier RA, Parker GD, Graham RI, Da Silva M, Halmagyi GM. 1997. Vestibular abnormalities in CHARGE association. Ann Otol Rhinol Laryngol 106:129–134.

Nicholas J. 2005. Can specific deficits in executive functioning explain the behavioral characteristics of CHARGE syndrome: A Case Study. Am J Med Genet (in press).

Pagon RA. 1999. The eyes in CHARGE for the ophthalmologist. In: Hefner M, Davenport SLH, editors. CHARGE syndrome: A management manual for parents. Columbia, MO: CHARGE Syndrome Foundation.

Salem-Hartshorne N, Jacob S. 2004. The prevalence of developmental delay in CHARGE Syndrome. J Early Interv 26:292–301.

Stjernholm C. 2003. Aspects of temporal bone anatomy and pathology in conjunction with cochlear implant surgery. Acta Radiol Suppl 430:2–15.

Stock Kranowitz C. 1998. The out of synch child: Recognizing and coping with sensory integration dysfunction. New York: Berkley Publishing Group, Skylight Press.

Thelin JW, Hartshorne TS, Hartshorne NS. 1999. Audiologic and educational issues in CHARGE syndrome. J Educ Audiol 7:34–41.

Weber BP, Dillo W, Dietrich B, Maneke I, Bertram B, Lenarz T. 1998. Pediatric cochlear implantation in cochlear malformations. Am J Otol 19:747–753.

Wilbarger P, Wilbarger JL. 1991. Sensory defensiveness In: Children aged 2–12. Santa Barbara, CA: Avanti Educational Programs.

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