

*This article is the first installment in a two-part series. Part Two will be published in an upcoming edition of reSources.*

## **Deaf-Blindness, Self-Regulation, and Availability for Learning: Some Thoughts on Educating Children with CHARGE Syndrome**

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Sigmund Freud claimed that there was no such thing as bad weather, only the wrong clothing (obviously they never get tornadoes in Vienna). In much the same way we can say that there is no such thing as a child with CHARGE syndrome who cannot be educated successfully in a local school, only programs which are not the right 'fit' for the child. Is it easy to get this right? Usually not, and it tends to become more of a challenge as the child grows older and moves into middle and high school, but there is evidence from a growing number of successful situations that demonstrates both the challenges and the attitudes and solutions that can lead to success. Reports from a large number of parents and education professionals over the past few decades confirm again and again what a challenge this can be. Often the biggest difficulties relate to behavioral features of the children, but these are usually compounded by inappropriate responses from the education world itself. The chief culprits that I see are lack of creative thinking, stereotypical and inflexible attitudes, inappropriate assessment procedures, a refusal or inability to genuinely observe children, and a misinterpretation of things that are observed. Attempts are made to force the children into a pre-existing, standard mold, often with behavior plans as a significant extra component, and the end result is almost always to intensify the problems and move even further from any desired educational outcomes.

People with CHARGE generally have to cope with much more than just functional vision and hearing problems because this condition often involves multiple systems and all, or most of, the senses. In my experience most people with CHARGE develop amazing adaptive behaviors and almost all of them end up "fooling" other people into thinking that everything is easier and more straightforward for them than it actually is. People who imply that a child's vestibular or vision or hearing issues have resolved and are no longer a problem have clearly misunderstood the compensatory skills that the child is having to use in order to function at their current level. A good and effective educational program, while being very positive and having high expectations, should always take account of the fact that everything that a child with CHARGE syndrome does is likely to take more thought and attention and concentration and energy and time for them than it does for those of us who work with them. The education establishment seems to be unduly fixated on, almost obsessed with, considerations of cognitive skills and potential to the exclusion of all else, yet for most children with CHARGE cognitive issues appear to be of lesser

significance compared to the overwhelming impact of multiple sensory and neurological issues that we are still struggling to identify and understand.

The key challenges are listed below:

### **Deaf-Blindness**

Most people with CHARGE that I have met satisfy the criteria for being considered as having deaf-blindness, even if they have some useful vision and hearing. This is a disability that is defined in functional, not clinical, terms and for each individual with CHARGE it is mostly about difficulties in accessing information not just from the world around them but even from their own bodies. In 2005 I wrote that,

“People 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.” (Brown 2005)

People who suggest that a child’s functional vision and/or hearing is fine inspite of the presence of features like ocular defects, persistent ear infections, very low muscle tone, poor tactile and proprioceptive perception, and significant vestibular dysfunction should be regarded with scepticism. If anyone wishes a broader view of vision issues in CHARGE syndrome, for example, and the complexities involved, go to [www.cadbs.org/newsletter/](http://www.cadbs.org/newsletter/) (Brown 2010).

The philosophy of deaf-blind education offers the best match for these multiple, complex, and overlapping sensory problems, as outlined in the article “Educational Needs of Children with CHARGE Syndrome” (Majors and Stelzer 2008). The list of teaching strategies at the end of this article gives a particularly good idea of the range of approaches that might need to be considered in order for the educational program to be successful. Once a child is regarded as having deaf-blindness a very special and appropriate world of resources and information is opened up. With the Internet, and websites such as those of the CHARGE Syndrome Foundation, DB-LINK, the Texas School for the Blind and Visually Impaired, California Deaf-Blind Services, and many others, advocates for this population now have a significant and growing body of information to assist them.

### **Curriculum Delivery**

For children with CHARGE it is particularly important to think carefully about ways that the curriculum is being delivered. Although a child might ‘perform’ well with an adult in quite structured activities, these activities could sometimes impose significant levels of stress if they continue for too long a period of time, or if the child’s positioning becomes uncomfortable or insecure. Because a child seems to have a good level of cognitive ability and potential there might be an assumption that they should therefore be able to sit for 20 or 30 or more minutes at a time and attend and learn in quite structured ways, rather like most other students of their

age with a hearing loss or a visual impairment. But cognitive ability/potential is only one of the things needed to learn and function in this kind of way, and in fact the child might have a range of disabilities, and especially sensory difficulties, that are likely to present big obstacles to managing these (for them) lengthy periods of immobile desk-learning.

An activity-based approach to curriculum (i.e., working on the appropriate curriculum goals in areas like math and language and social and self-help skills, but all embedded within certain functional activities that require the child to move around periodically) can often help to keep stress levels and arousal levels down. Many children with CHARGE enjoy and respond well to moving around and physically ‘doing’ things, and can learn a lot of the curriculum while they are doing this. They also, and paradoxically, often need to get into a fully-supported horizontal position, or at least place their head in this position for periods of time. Of course, a certain level of ‘desk-learning’ should be introduced as a part of each day also. Keeping these sessions fairly brief, requiring the child to move when they seem to be losing concentration, and using materials and concepts that are motivating and meaningful to them should help to guarantee a fair degree of success. Sometimes the child might need lots of time to gather information, work at things and get them right, but at other times they might need activities that do not last too long and that move at a good brisk pace; otherwise they might become stressed or bored and may need some kind of specific sensory input. In particular, people need to learn how to ‘read’ the child to see when they are becoming over-aroused, or stressed, or tired, and so are needing to be helped to transition smoothly to a more appropriate activity.

### **Communication & Language**

There are many important and complex considerations in the area of communication and language. For a variety of reasons, many children with CHARGE have difficulties with accessing purely spoken language, especially with using it themselves for expression. But they do show great interest and skill in receiving information visually (i.e., signs, gestures, familiar objects) and in expressing themselves gesturally. While it might be important to continue using appropriate spoken language with the child, I think it is important for most children in this population to offer them language in sign and to think about using gestures, facial expressions, and objects communicatively as well, and going for a Total Communication approach (see Majors & Seltzer). If the child does later opt for speech as their expressive language mode, there may be several specific reasons why they have some difficulties with the articulation of speech (see the list below), and giving the child access to sign as a backup for their expressive speech should also help to reduce any potential frustration levels at not being immediately understood by other people. The child may well then make their own choices and develop their own preferences about how and where and when and with whom they use speech or sign. Many children, if they have a range of communication options available to them, both receptively and expressively, clearly show their skill at choosing what suits them best from moment to moment. It is not at all unusual for a person with CHARGE syndrome to develop good levels of language but to receive in one mode (often speech) and to express in a different one (often sign). There is also evidence of gradual or sudden deterioration of hearing and/or visual abilities in some people with CHARGE, which adds further weight to the argument that Total Communication is a wise option to maintain.

One additional point that needs to be mentioned is the growing likelihood that many people with CHARGE might have executive function disorder (see below). This carries many implications, but as far as communication is concerned it may make it difficult for children to initiate it, so that even if they have the language (spoken or signed), and even if they know exactly what they want or need to say, they might be unable to do it unless another person gets them started (e.g., by asking the child a question like “Are you okay?”, or “Do you want something?”, or “Do you need to go to the bathroom?”). Sometimes, a tool like a written or pictorial daily calendar system might be appropriate and helpful, along with some kind of individualized communication book that includes a vocabulary of relevant written words or drawings of signs, which the child can refer to to help get themselves started. These concrete systems might offer children an important amount of support and reassurance even though they may not obviously refer back to the calendar often throughout the day. My experience suggests that many students with CHARGE benefit from simply knowing the calendar and/or the communication book is there and can be consulted if needed.

### **Potential Barriers to Speech & Sign Articulation**

When we consider the many anomalies that are associated with CHARGE syndrome we find many that are likely to have a negative impact on the development of well-articulated speech or signing. I will list these significant anomalies first from the perspective of barriers to effective speech articulation:

- Hearing impairment
- Vision impairment
- Facial palsy
- Low muscle tone
- Poor tactile perception
- Oro-facial clefting
- Enlarged tongue
- Poor tongue movement
- Small lower jaw
- Larynx/pharynx anomalies
- Breathing difficulties and poor lung capacity
- Swallowing difficulties
- Dental anomalies
- Delayed/immature eating skills
- Chronic congestion/infections

In spite of this daunting list many children with CHARGE do opt for speech as their preferred expressive mode, although articulation difficulties may make their speech very difficult to understand. The list of anomalies that may have a negative impact on good sign articulation is just as imposing, although generally speaking it is easier for children to form intelligible signs than intelligible words. Here is the second list, from the perspective of barriers to effective sign articulation:

- Low or poorly modulated muscle tone
- Poor tactile perception
- Poor proprioceptive perception
- Poor vestibular perception
- Poor spatial awareness
- Dyspraxia
- Poor body awareness
- Poor bilateral coordination
- Poor sense of equilibrium
- Various postural difficulties
- Low vision, blindness or visual field losses
- Skeletal anomalies

### **Avoid Working at Thresholds**

I think of most children with CHARGE syndrome as a people who, for much of the school day, are probably operating at the very thresholds of their sensory abilities, which is a challenging and potentially stressful level at which to be functioning for more than short periods of time. This is possibly the situation even at times when the children seem quite relaxed and confident and the activities seem well within their various competencies. Consequently it is a good idea to try to ensure that activities are simplified and made as easy as possible for each child so that the essential teaching point is clear and accessible and the child is not having to think about and prioritize other considerations (e.g., dealing with background noise, or challenging physical positions, or visual clutter or glare, or other kinds of sensory overloading, or anxiety about unknown future plans, and so on). The children's level of alertness or fatigue appears to show itself in their posture and movement patterns and attention levels, and people involved with them need to familiarize themselves with these indicators and know how to adapt each situation so that the child is supported and does not become too tired or too stressed.

### **Complex Health Issues**

These are typically complex and severe in nature, and often and repeatedly are life-threatening in nature, so they take priority over educational considerations. Thus there are significant considerations around the availability of medical/para-medical services, and the strategies required to balance health and educational needs. In the broad special education field there is extensive literature about these kinds of issues in school settings. It is not so very rare to see a child with CHARGE in school with a full-time nurse, maybe also with a full-time educational intervener or a sign interpreter, and in my experience this has always seemed necessary and appropriate in each case I have observed. If nursing and healthcare procedures take up much of the school day that is where the education program can focus. So much of the curriculum can still be accessed during these procedures which are not an excuse for abandoning any idea of dialogue, communication and language development, numeracy and literacy work, orientation and mobility activity, turn-taking, for example. In this population early vocabulary will often include the words/signs for such things as ventilator, G-tube button, suction, pills, and so on. It

may also be possible to work towards independence with some of these nursing/para-medical procedures; it seems to me quite iconic and typical that the first child I ever saw give himself lunch through a G-tube entirely independently (while he continued to converse in sign language one-handed) should have been a teenager with CHARGE!

### **Executive Function Disorder & Self-Regulation Issues**

With the passage of time I have come to consider this as probably the most challenging and least understood long-term aspect of this condition for the children themselves, their families and educators. Whenever I am approached and asked to help with a student with CHARGE it is almost always these issues that are the main focus of the challenge and so of the solutions. Several people in the field are now investigating, among them Tim Hartshorne who wrote this in the CHARGE Syndrome Foundation Professional Packet:

“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.” (Hartshorne 2005)

So many issues encountered in a significant portion of the population of people with CHARGE – poor self-regulation, rigid dependence on routines, difficulties managing transitions, maintaining an appropriate emotional perspective, urgent but unusual postural needs, sensory needs and resultant self-stimulation behaviors - all add layers to the puzzle. The first step seems to be to try to ascertain when these issues, and which of them, are present before anything effective can be done about them. My observations suggest that most children with CHARGE are not always and not fully available for learning due to this multi-layered set of issues, and that this needs to be considered urgently before other aspects of the academic curriculum can be addressed effectively. We are learning from examples of creative thinking and good practice, much of it crisis-driven. The use of interveners (see <http://intervener.org/> for example), an idea that comes from the field of deaf-blind education, has proved very effective in many cases – in aiding more efficient ongoing assessment, providing more effective access to the curriculum in fulfilment of IEP requirements, in facilitating interactions with peers and an ever-growing social circle, and in offering reassurance and effective strategies when self-regulation issues begin to arise. We still have a long way to go, but at least the focus now seems to be more clear and more appropriate.



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