
**“Why are children with CHARGE syndrome so lazy?”
Reflections on caution, self-preservation, adaptive abilities,
function, efficient use of energy, and self-awareness, and the way
that these can be misinterpreted.**

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Children with CHARGE syndrome frequently seem to be uniquely driven and goal-oriented, full of energy, curious, narrowly focussed and insistent on getting what they want, and unwilling to compromise. These characteristics can be observed even in infants who have not yet acquired any independent mobility, as well as in older children. Even though children with CHARGE can be lazy, the way all of us can be at times, “lazy” is just about the last word I would use in any description of them, so it surprises me how often I hear the word “lazy” used when people talk about them. The word usually crops up in connection with the fact that a child is not yet walking independently even though it looks as if they could, or sometimes because they often like to stretch themselves out in the horizontal position, or they rest the side of their head on the table for periods of time. The word “lazy” is almost the most commonly used adjective for children with CHARGE (“naughty” seems to be the most popular, but then it would be, wouldn’t it?) yet in my experience the children are almost all determined to get up and go when the time is right, to such an extent that I don’t understand why they are not much more lazy than they actually seem to be. How many parents have felt as if their child with CHARGE was never ever going to walk, then one day at the age of three or five or eight the child has just, apparently quite suddenly, launched themselves into space and become a walker? Readiness is all!

Very few professionals get any training about vestibular issues, and they get no training at all about CHARGE syndrome of course, so that often their rather negative comments might simply come from them being unaware of all the complexities involved for the child. It seems like a good idea to try to clarify what some of these characteristic behaviors might mean, and what they result from, so that the “I” word can be removed from discussions of development and behavior.

Standing and walking

The challenge of developing good equilibrium (balance) and postural security for independent standing and walking for children with CHARGE is an outcome of many different aspects of the syndrome including visual problems, deafness, vestibular issues, low muscle tone, loose

connective tissue (the tendons and ligaments that bind the joints), skeletal issues, poor proprioceptive perception, breathing problems, poor body awareness, difficulties with executive attention, and who knows what else. Let's take a brief look at some of these considerations in turn.

The Equilibrium Triad

Although the vestibular sense is often called "the balance sense," balance actually develops from the interaction of vestibular input with proprioceptive input and visual input. Since all three of these senses are usually missing or impaired in children with CHARGE it should be no surprise that independent standing and walking develop only very slowly. And while that development results from growing experience, and strength, and confidence, it also develops from a host of smart adaptive strategies that the children have to learn to use in order to compensate for what is missing from each of these three crucially important sensory systems. The references at the end of this article will all touch on these aspects of development in more detail.

Vestibular Issues

These are probably the biggest and most significant obstacles for children with CHARGE to overcome and alone could explain why walking often develops quite late. One problem with developing effective standing and walking is that most children devise their own adapted ways of moving from one place to another to minimize the impact of their vestibular deficits and the challenge of resisting gravity. Movement strategies like the five-point crawl, the back scoot, side-winding, and rolling, become so safe, so quick, and so effective that risking the catastrophe inherent in standing and moving on only the two feet would be a foolish choice for the child to make until they know that they are absolutely ready for it. Maryann Girardi once told me that for children with CHARGE "gravity sucks," and we would do well to remember that when the child seems stubborn or fearful or resistant to our efforts to get them walking.

Vision Problems

A child with missing vestibular sense and poor proprioception might eventually look as if their balance is quite good when they are standing, and especially when they are walking, but in fact they might be using their vision to fixate on a distant object to guide them in keeping upright and moving on target as they walk. If this is the case the child does need to maintain their gaze on that target object for the entire journey until they arrive. If they get distracted and take their visual attention away from the target during the journey they are very likely to veer off course, or even collapse to the floor like a tree being felled. The biggest problem with this over-reliance on visual input is that it doesn't work if there are no clear visual objects around on which to focus the eyes, it doesn't work in bright light with lots of glare (especially if the child has iris colobomas), it doesn't work in low light or darkness, and it prevents the eyes from moving around during the journey (known as ocular motility) to check for obstacles on the way, especially down at floor level and in the left and right peripheral visual field. Visual field loss in CHARGE is almost always in the upper visual field, so when a child regularly trips over obstacles on the floor people get confused and assume that there must be lower visual field loss as well. In fact the lower visual field might be fine, but the child cannot make their vision available for checking the floor because their vision is serving the much more urgent need to keep them

upright and moving in the right direction. As a result of all these visual factors a child might walk independently at certain times in certain places, but revert to back scooting or some other safer and easier method of moving at other times and in other places.

Breathing Difficulties

In her three-part webinar PT Dinah Reilly makes many important and relevant points even though she is not speaking specifically about CHARGE syndrome. One of the things I liked most about her talks is when she mentions the way that difficulties with breathing get in the way of good postural control and balance. She quotes from an article by Hodges:

“Every muscle in the trunk is both respiratory and postural muscle, especially the diaphragm. If breathing is compromised, postural muscle activation is reduced to focus on immediate needs of respiration and thus posture is de-stabilized.”

This quote also reminds me of Jean Ayres’ comment that after air to breathe, postural security is our next most urgent priority. Given the high frequency of breathing issues in the population of children with CHARGE this connection needs to be remembered and recognized when the focus is on the development of walking.

Challenges for the Physical Therapist

Maryann Girardi, at Perkins School for the Blind, has made a very interesting short webcast called “CHARGE Syndrome: Providing Physical Therapy.” She speaks from a long experience of working with children with CHARGE, and perhaps the most interesting thing about her presentation is that she spends very little time on conventional PT skills and techniques. Instead she talks about things like the importance of building a positive relationship with the child, being consistent and predictable, communicating effectively with the child, using the child’s motivators and setting realistic goals for the session, observing to ensure that the child is in a good level of arousal (not over-excited or drowsy), and allowing the time necessary for the child to be actively involved in what is going on. Her concern is not with getting the PT right but with creating a situation where her PT skills can be delivered effectively and involve the child.

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