THE INFLUENCE OF SENSORY LOSS ON DEVELOPMENT

See also the Manual sections on Development

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INPUT IMPAIRMENT

In terms of intellectual development, the most important anomalies in CHARGE involve the eyes and ears. Vision and hearing are the most important "distance" senses. Some people have estimated that 70% of what we learn is through our vision and yet language develops most quickly with hearing unless the child is born into a signing environment. The combination of hearing and vision loss has been called "dual sensory impairment" or, more simply, "deafblindness." I also like to use the term "input impaired." Most measures of intelligence rely on output, i.e. how well the children use their hands, walk, talk, etc. From these outputs, an estimate of mental processing is reached. However, if input is inadequate, how can processing and output be optimal? As a result, the intelligence of children who are deafblind is routinely underestimated. While some children with CHARGE may have brain malformations which limit intelligence, *I believe the majority of these children have normal or near-normal abilities to process input*. However, that input has to be presented in a manner that the children can understand.

DETERMINING THE COMMUNICATION BUBBLE

Programs for those individuals who are blind/visually impaired usually count on hearing as a compensatory skill whereas those for the deaf/hard-of-hearing use vision. What happens if both are limited? Different approaches to learning must be found. Finding a program with at least some staff who understand dual sensory loss will be vital for the optimal development of these children. These staff can then teach those with more traditional backgrounds to adapt their programs. Using the sensory inventory to be published in a manual for interveners[1] can be useful for families and educators in determining the "communication bubble." This bubble is the area within which a child can comfortably communicate with another person or get input from the environment. The area will be different for each of the five senses: vision, hearing, smell, touch and taste.

VISION (also see section on Eyes)

If a coloboma is present just behind the iris, that section of retina will not function properly, creating a blind spot in the child's vision above the eyes. This is normally small and does not interfere much with seeing. If the cleft is large and involves a significant portion of the lower retina, then the blind spot may also be large. That child will not be able to see anything above the level, for instance, of the top of the eyeglasses. He/she will have to look up or tilt his/her head up in order to see what is up there. More significant is a cleft involving either the optic disk, which is the nerve coming into the eyeball, or the macula, which controls central vision. In either of these cases, the child may not be able to see objects clearly and distinctly no matter what kind of glasses are used.

Many children with CHARGE have different vision in each eye. In addition, some of them may have amblyopia. This occurs when the vision in one eye is better than in the other or when the eye muscles are weak on one side so the child cannot focus well using both eyes. The brain suppresses the image from the poorer eye so they do not have to contend with double vision. Sometimes the children have eyes that are of unequal size as well. A smaller eye usually means that the cleft inside is larger or more significant; the vision on that side is usually worse.

The effect of these anomalies is an upper visual field cut and/or uncorrectable blurry vision. Such a visual field cut means the child will bump into object above him/her such as tables or open cupboard doors, or will have to tip the head back in order to look up at an adult (this can be hazardous if balance is not good).

If the macula is involved, the central vision may be very blurry or even absent. With central vision loss, the child will look above the target. For instance, in order to see a person's eyes, he/she would look at the forehead or hairline. Such children may be labeled autistic because they do not appear to make eye contact in addition to having poor communication skills because of deafness.

A *vision consultant* can help adapt materials to fit the vision needs. An *orientation and mobility specialist* may be needed to help the child move safely about the environment.

HEARING - Please see the section on Hearing in CHARGE for more details.

The hearing losses in CHARGE can be anything from mild to profound. Most children with profound hearing loss, however, do have better residual hearing than is recognized during the early hearing evaluations. Recent evidence shows that any kind of language program (oral/aural or sign) initiated for a deaf/hard-of-hearing infant within the first six months of life can lead to significantly improved language development.[2] This is clinical confirmation of the basic science evidence that the brain pathways for hearing (as well as other sensory modalities) do not develop if the input is not presented while the brain is developing.

BALANCE

When the vestibule of the inner ear is involved, the children are born without the balance sense that comes from the inner ear. This balance sense tells the child where his/her head is in space: up down, tipped sideways, or in a diagonal direction. When this sense is not present at birth, the child feels unsteady as he/she raises the head. If that same child does not see very well, there is very little motivation to raise the head. Therefore, these children have very delayed gross motor development. They prefer to keep their bodies flat on the ground or in a stable condition. They may roll to where they want to be, combat crawl, crawl with their head down on the ground (a 5-point crawl), or sit and bounce forward or backwards to where they want to be. When they get up and walk, they hang on for longer periods of time and they also keep their feet wider apart. When both vision and hearing are affected, the average age of walking in CHARGE is 3-4 years. Unsteadiness may persist into adulthood if vision is significantly impaired. The CHARGE Syndrome Foundation has a videotape of children with inner ear balance problems which shows gross motor development from infants to young adults. Children learn to trust their muscles and joints and do learn to walk and run, though on a delayed schedule.

TONE

Many children are also reported to have low muscle tone (hypotonia). In some cases, the upper body is more involved than the lower body. This has not been well studied. However, we think that the muscles themselves are normal and that the major problem is muscle control from the nervous system.

OTHER SENSES

Touch becomes the major means of input if vision and hearing are limited. This can be with the hands, feet (preferred in some cases), face and tongue. Young children routinely mouth objects. This becomes socially unacceptable in older children and, yet, mouthing objects is a legitimate way of getting further information when other input is not available. Most children with CHARGE have a good sense of light touch. Pain sensation, however, is often altered. Many have very high thresholds for pain. Since they do not experience pain normally, they may inflict pain on others without realizing what that pain is. This can become a social problem.

Smell is frequently diminished or absent in CHARGE. The consequences are important for nutrition since smell constitutes the major part of flavor. When your nose is stuffed up from a cold, food does not taste as good. In addition, children may not understand why others move away from them when they pass gas or, as teenagers, when they take off their shoes or have just come in from vigorous exercise. They simply do not smell what others do.

Taste is probably normal but this has not been investigated. Taste buds detect only four qualities: salty and some sweet on the front of the tongue, sour on the sides and bitter in the back. Therefore, children without sense of smell may be more likely to prefer salty and spicy foods. Interestingly sweetness is not as prominent when smell is absent.

OTHER FACTORS

The child who has spent many days, weeks, or even years in the hospital does not have the normal experiences developmentally. If he/she cannot hear well, see well, or balance properly, then all phases of development are delayed. This includes gross motor, language, personal-social skills, and even fine motor skills. Understanding what is happening in the eyes and ears is, therefore, a very important task, especially in the first year of life. This rarely happens, however, because the doctors and families are more concerned with the structural problems with the nose, the palate, the heart, the kidneys, the esophagus, etc. Filling out a Developmental Timeline can be helpful in showing the interaction of the many medical concerns with developmental milestones.[3]

SUMMARY

In the long run, the measure of intelligence and the ability to develop into an independent and contributing adult depends on the level of communication that can be achieved. If a child cannot tell you how he/she thinks, you cannot measure his/her intelligence. Routinely, therefore, tests of intelligence conclude that children with CHARGE are retarded. In some cases, certainly, children have difficulty with cognition and could be called mentally retarded. In most cases, however, it is my contention that children are developmentally delayed to a very significant degree, but we cannot accurately measure what their cognition is. As we learn more

about adolescents and young adults, it appears that some of them have significant learning disabilities. Exactly what these learning disabilities are is just now being studied.

It is important to understand the interaction of the multiple anomalies present in CHARGE. From a functional point of view, it is crucial to find a way of establishing a formal communication system, both at home and at school, so these children can reach their maximum potentials.

REFERENCES

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