

## MEDICAL INFLUENCES ON DEVELOPMENT IN CHARGE

Sandra L.H. Davenport, M.D. SensoryGenetic/Neuro-development  
5801 Southwood Drive, Bloomington MN 55437-1739  
952-831-5522 [slhdaven@tc.umn.edu](mailto:slhdaven@tc.umn.edu)

Medical problems and overall health have a tremendous impact on development. One of the things which makes CHARGE syndrome so complex is that each medical feature can be 0% (absent) to 100% (severe involvement) in any given child. The purpose of this table is to provide a quick visual reference for most of the medical issues the child and parents have to deal with. Features on the severe end of the spectrum should be viewed as priorities. However, mild problems should not be ignored, as they may significantly complicate the overall picture.

On the back of this page is a summary table listing functional problems that have an impact on learning. You may wish to check off those problems which are present (left most column) and make a mark on the scale on the right indicating mild to severe involvement for each feature in your child. Don't get hung up on the percentage. This is only a gross estimate, the purpose of which is to know the relative impact of each when compared to the whole.

Y N	System Affected	Condition	0%-----50%-----100%		
			Mild	Moderate	Severe
	Eyes	Vision loss (coloboma)			
	Ears	Hearing loss			
	CN* 1	Lack of smell			
	Choanae	Nose breathing difficult, stents			
	Inner ear	Balance problems			
	Receptors	High pain tolerance			
	Receptors	Tactile defensiveness			
	CN 7	Facial palsy Rt, Lft, Both			
	CN 9&10	Swallowing problems			
	CN 9&10	Recurrent aspiration			
	CN 9&10	Tube feeding			
	CN 9&10	Tracheostomy care			
	CN 9&10	Copious secretions			
	Heart	Heart problems			
	Kidney	Reflux, kidney probs			
	Growth^	Small for age			
	?Immune	Ear infections			
	?Immune	Sinus infections			
	?Muscles	Hypotonia			

\*CN = cranial nerves which supply the head and neck. All 12 may be affected but the most common are listed.

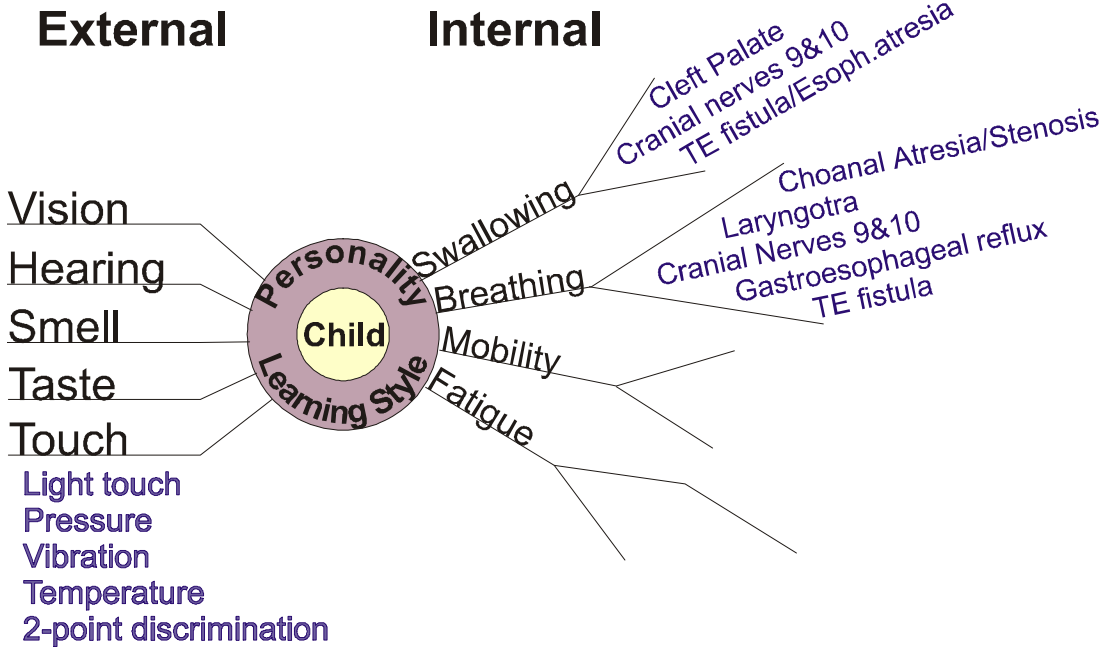
^Some children have problems with growth hormones but more are small because their bodies use up so much energy from illness or because they do not get enough nutrition due to the swallowing problems.

# PHYSICAL INFLUENCES ON DEVELOPMENT IN CHARGE

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By Sandra L.H. Davenport, M.D.

CHARGE is such a complex disorder that many of the physical components have a profound effect on the child's development and understanding of the world. The above diagram breaks these influences down into those that are External and those that are Internal. In one way all are internal since the five senses are all part of the body; however, these senses require outside stimuli in order to perform. Pain is placed between the two because it can occur because of both external and internal events.

The Internal influences that have the most profound effect are problems with swallowing, problems with breathing, delayed mobility and problems with fatigued and illness. Each of these problems has multiple causes, which are listed and further explained in the medical section of the manual.

The "External" senses are the source of contact with the outside world. Decreased vision impairs understanding of the environment. The vision loss in CHARGE is somewhat unique because most children have colobomas, which cause upper visual field losses. This means that they may have blind spots above the direct line of gaze. More importantly, they may involve the macula or the optic nerve, which will cause blurred vision.

Decreased hearing results from malformations of the middle and inner ears and/or middle ear infections. Hearing loss is common in CHARGE and causes major problems with understanding and imitating speech as well as not being the able to detect environmental sounds. Most children that have hearing loss due to malformations of the inner ear also have problems with balance because the vestibular system is malformed.

Smell may be decreased or absent in CHARGE. This results in lesser appreciation of food and in social mishaps because they don't know when they stink. Smell is also important because it is the third distance sense. They can often tell who is coming by perfume or body odor. Rooms of the house in different places outdoors may also have distinctive smells. These may help them figure out where they are.

Taste is almost always normal in children with CHARGE. It can be important for exploration when the other three senses are not working. Older children who have profound vision and hearing loss and have not had adequate communication training may spend a lot of time mouthing objects. Putting objects in the mouth gives more information than simply handling them.

Touch is actually much more complex than people realize. The sensors under the skin can distinguish between sharp and dull, soft touch and pressure, vibration, as well as differences in temperature. Two-point discrimination means being able to tell the difference between one pin and two pins. The pins can be very close together on fingertips and still recognized as two objects. Most of these aspects of touch seem to be normal. However, a lot of children prefer deep pressure to light touch.

Pain may be different in many children with CHARGE compared to their peers. Parents report that their children have very high pain thresholds. One consequence of this may be that they cannot understand why other people react to painful pinches or kicks etc. Because they don't feel the pain themselves they may not be able to empathize with other people's pain.

## INFLUENCE OF SENSORY LOSS ON DEVELOPMENT: The Communication Bubble

Sandra L.H. Davenport, M.D. Sandra L.H. Davenport, M.D.  
SensoryGenetic/Neuro-development  
5801 Southwood Drive, Bloomington MN 55437-1739  
952-831-5522 [slhdaven@tc.umn.edu](mailto:slhdaven@tc.umn.edu)

### Are all five major senses working?

The chart below is not based on any reliable data. Estimates are quoted in the literature that the percentage of information that we take in via our eyes is anywhere from 60 to 90%. Of course, if you are listening to an audio tape, vision doesn't matter at all. If you are watching a film strip, hearing is of no consequence. In addition, some people are visual learners and some people are auditory learners. It is well-known that two people with identical audiograms may have strikingly different abilities to understand speech and other sounds. Perhaps the one who understands speech better is actually an auditory learner. But even that person misses information. If he/she uses speech-reading extensively, then a vision impairment on top of a hearing impairment will cut down on understanding as well.

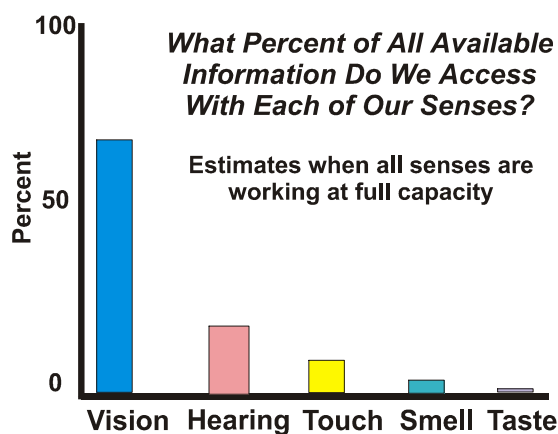
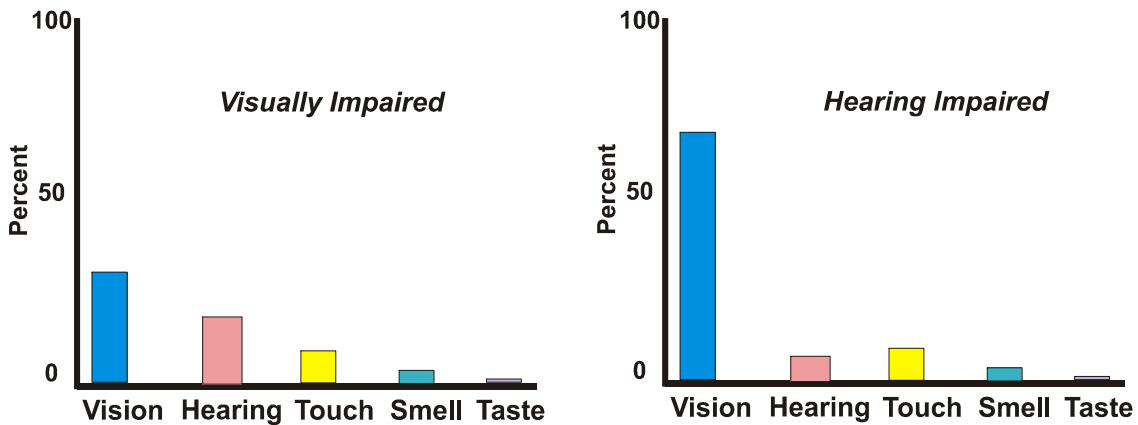


Figure 1: Full access to information from all senses.

However, consider what happens if a child is visually impaired AND hearing impaired. If a child has moderate visual impairment, the Vision bar might be half as high. If the child is moderately hard of hearing, the Hearing bar would be shortened by half. Relatively speaking, then, the other senses become more important.



Figures 2 & 3: Compare the differences when half of either vision or hearing is gone.

Now consider how less information is available when half of BOTH the vision and the hearing are missing. Notice how much more important the senses of touch and smell become.

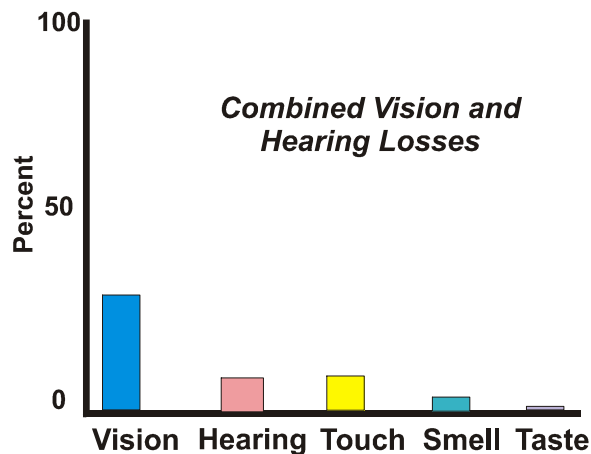


Figure 4: Combined vision/hearing loss with half of each gone.

## **What is Deaf-Blind?**

**Definition:** Any combination of hearing and vision loss that interferes with access to communication and the environment and requires interventions beyond those necessary for hearing or vision loss alone. For educational purposes in most states, the child needs to meet the criteria for deaf/hard-of-hearing as well as for blind/visually impaired (check with your own DeafBlind Project director)

Deaf-Blind rarely means totally deaf and totally blind. No other succinct term has been found that fits all of the conditions listed in the table below. Dual sensory loss, Hard of Hearing/Visually Impaired, etc. are cumbersome. Most parents and educators have finally settled on DeafBlind even though that term is a somewhat inaccurate and, often, a scary term.

The importance of having a DeafBlind label, however, cannot be underestimated. The educational needs are truly different when both senses are affected. It is not enough to have the consultants come in and give their input without considering the effect of the other sensory loss. The reason is that the techniques used to compensate for vision loss often involve hearing and those used for hearing loss frequently involve vision. Note in the table below that the term DeafBlind is used even if the child is found to be just visually impaired and hearing impaired. The combination makes a HUGE difference in the way information should be presented.

## **What combination of vision & hearing is present?**

When one or more senses are impaired, additional educational consultants are needed, even (or perhaps especially) during the Early Childhood phase of development. Research out of Denver shows that, if a communication system is established BEFORE six months of age, a deaf child can develop completely normal language. Even if they learn to read sign language, they do not fall behind their hearing peers in expressive language and reading. The implications are staggering. Even though medical issues in CHARGE occupy the thoughts and minds of parents and caretakers during the first 2 years of life, it would appear to be very important to start some form of communication as early as possible.

The table below introduces the kinds of special educational consultants needed to address the vision and hearing issues. However, physical therapists, occupational therapists and a variety of other consultants may also be needed (just like in the hospital and clinic). Note that the kinds of modifications given depend on the nature of sensory input.

<b>Sensory Status</b>	<b>Consultant Needed</b>	<b>Modifications</b>	<b>Comments</b>
Hearing Sighted	None		
Hearing Visually impaired	B/VI teacher +/- O& M	Large print, contrast, placement of student	
Hearing Blind	B/VI teacher +O& M	Braille, O&M, audio tapes & instruction	
Hard of Hearing Sighted	D/HH teacher +/-SPL	Hearing aids, +/- FM or other amplification, noise reduction, classroom placement	
Deaf Sighted	D/HH teacher +/- Interpreter	Deaf classroom or program, interpreters	
Hard of Hearing Visually Impaired	B/VI, O&M, D/HH, SPL, DBP consultant	Amplification, large print, be within the "Communication Bubble"	Deaf-Blind
Hard of Hearing Blind	B/VI, O&M, D/HH, SPL, DBP consultant	Amplification, Braille, O&M, +/- sign language	Deaf-Blind
Deaf Visually Impaired	B/VI, O&M, D/HH, DBP consultant	Modified sign language, O&M, large print, be within the "Communication Bubble"	Deaf-Blind
Deaf Blind	B/VI, O&M, D/HH, DBP consultant	Modified sign language (possibly tactile), Braille, O&M	Deaf-Blind,

Table modified from Rob Last

B/VI = Blind/Visually Impaired

D/HH = Deaf/Hard of Hearing

SPL = Speech/Language

O&M = Orientation & Mobility (learning how to know where you are and move safely with low or no vision)

DBP = DeafBlind Project



## The Need to Establish A Communication Bubble

Establishing what Susan Smith, the parent of two children with CHARGE, calls the “Communication Bubble” is essential if you want to make sure the child knows you are there and are trying to communicate. The concept of a “bubble” is a good one because you can imagine the space within which you need to be. As Eric Kloos says, you need to be “on the child’s radar.” Get outside the bubble and you might as well not exist. Too many times, we think the child is tuned out, not paying attention, or is too “retarded” to answer when, in fact, he/she may not even know that you are trying to say something.

Each parent should know how far a child can see what kinds and colors of objects, with or without movement, in different lighting conditions and against different shaded backgrounds. The eye doctor will usually not be able to tell you this except by guessing though some do this kind of testing. A vision teacher, however, can do a Functional Vision Evaluation, to help establish these visual abilities. Figuring out what the child hears clearly, at what distance and on which side is also important. Neither of these is easy to determine so parents and team members will likely have to engage in a continuing evaluation process.

## What Types Of Communication Systems Are Available?

Emerging language	Touch cues	
	Object cues	
	Gestures	
	Pictures	
Formal language	Oral Language	Auditory/Verbal
	Oral Language supported manually	Cued Speech
	Sign Language	Signed English
		ASL, Auslan, etc. (the native sign language of a given country)
Total Communication	Combination of oral and sign languages	

Table modified from Rob Last

## How Do You Choose the Right Method?

There is no one right answer to this question. Too often, however, the confusion results from passionate arguments for one method of communication, like spoken English or American Sign Language, without fully understanding the effects of dual sensory loss. **The most important thing is to get language into that developing brain in a form that can be consistent and effective.** What that form takes will likely be determined a team meeting of the parents and professionals. We learn any language by receiving it and then repeating it. We learn it fastest and best by total immersion. Learning it for an hour in the classroom twice a week or in bits and snatches with a word or sign here or there is simply not enough to become fluent. This means that whatever method is chosen needs to be woven into everything the child does, from getting up in the morning to going to bed at night. Keep in mind that perfectly good language by whatever method is useless to the child unless it is delivered within the communication bubble. The other very important point is that a lot of communication precedes formal language. Every mother can “read” her child’s communication regardless of whether that child uses any formal

language. Sometimes it is guessing, but the child will let the mother know when she's got it right. Therefore, it is important to lead up to formal communication step by step. We use sight cues and verbal cues like outstretched arms with some encouraging words to let a hearing/sighted child know what we want to do next. For a child without good vision or hearing, concrete object cues and touch cues are used instead. DeafBlind project staff can help the regular and special education staff learn how to use these methods. As parents and educators we may get hung up on wanting our children to speak our own language using the method we use. That is as natural as immigrants wanting their children to continue speaking their language and remembering their culture. However, the issue is not the form but the substance. The child needs to learn that an object, touch, picture, word or sign is a symbol that stands for a thought. Many of them strung together convey complex ideas or stories. They include naming of objects, actions, remembering the past, anticipating the future and, later, getting into abstract thoughts and discussions.

So, above all, just get to it! COMMUNICATE, COMMUNICATE, COMMUNICATE.

# BEHAVIOR AS COMMUNICATION

Tim Hartshorne, Ph.D.  
Department of Psychology, Central Michigan University  
Mt. Pleasant, MI 48859  
(517)774-1779 phone (517)774-2553 fax [tim.hartshorne@cmich.edu](mailto:tim.hartshorne@cmich.edu)

All children behave. Much of this behavior is a matter of concern to parents, teachers, and others. Bookstores are full of information on how to cope with the misbehavior of children. No one should be surprised that children with CHARGE also have behavior that is a matter of concern. While the misbehavior of any child can be challenging, the misbehavior of children with CHARGE can be overwhelming for several reasons. First, there are frequently communication problems that make it difficult to discuss behavior problems and expectations with the child. Second, medical complications may limit the kinds of responses to misbehavior that parents and others are willing to utilize. Third, some of the behavior demonstrated is quite unusual when compared with the behaviors of the typical child.

When considering behavior management, one needs to be very clear about its goals. Eliminating misbehavior is not achievable, and by itself is most likely not desirable. To understand this, two important principles need to be described. First: all behavior has a purpose. Behavior is not random. Sometimes the purpose may be very simplistic, like scratching an itch on my head, but it is goal oriented. This means that the behavior demonstrated by our children with CHARGE is not random. They engage in it for a reason. The second principle is equally important: behavior serves a communicative function. This means that we use behavior to communicate something to other people. If we understand the communication, and if we understand the purpose of the behavior, then any behavior can make sense.

Now, to return to the issue of eliminating misbehavior. If we focus our efforts on stopping a behavior, we have taken away a means to a goal, and a means to communicate. That may suggest to the child that we are not interested in their goals or in communication with them. This may lead to withdrawal on the part of the child as they give up trying to engage us in communication, and it may lead to different, perhaps even more difficult, behaviors to try to achieve what the child experiences as important goals.

Thus the primary objective of behavior management is to understand the purpose and communication underlying misbehavior, and to use that to help the child find more appropriate communication methods to achieve their goals, or to help them find more appropriate goals. A temper tantrum may be a means to communicate the desire to receive attention, but children also have to learn that they need not be the center of attention all of the time.

When considering the goals of behavior management, six major principles need to be considered. **First**, as already stated, problem behavior serves a purpose for the person displaying it, part of which is communicative. Communication in general is fraught with difficulties. That is because all communication is subject to encoding of messages, and then decoding and interpretation. If I choose to communicate my frustration about a situation, I must first take my message of frustration and put it into some kind of communicative format, usually language. This is encoding. The listener must then hear my message and interpret it. This is decoding. Of course the listener interprets based on their frame of reference at the moment.

People with outstanding communication skills are articulate in encoding their messages, and accurate in decoding. However, when dealing with a person who lacks communication skills, and who may in fact be nonverbal, communication becomes extremely difficult. That is why communication with children can be a problem, and certainly why communication with children who have CHARGE is difficult.

**Second**, functional assessment is used to identify the purpose of problem behavior. If you know the purpose, the behavior is understandable. Our first task in working with a child with behavior problems is to understand the message behind their communication. We must decode their encoded message. To do this we try to determine the function or purpose of the behavior. Much has and is being written about methods for functional assessment. A great deal of training is needed to utilize some of them. IDEA requires functional assessment in the schools, and school psychologists should be able to conduct such an assessment.

**Third**, the goal of intervention is education, not simply behavior reduction. Once you believe you understand the purpose of the behavior, how do you intervene? The difficulty here is to modify the misbehavior without shutting down the child's attempts to communicate. Here are several general considerations. First, do not become annoyed, threatened, hurt, or give up. Second, help the child find a more appropriate method of communicating the message to you. A third consideration is to avoid crisis management. This involves ignoring the behavior, protecting the individual and others, restraining the individual if necessary, removing anyone who is in danger, and engaging in behaviors that generally reduce the misbehavior. The problem with crisis management, which of course must be used at times, is that it does not teach the child new ways of communicating and reaching their goals. A fourth consideration is to be patient. It takes time to modify behavior in a way that teaches the child the benefits of clearer communication.

**Fourth**, problem behavior may serve many purposes and therefore requires many interventions. The same behavior, for example having a temper tantrum, may be used when the child wants something, and when they want to avoid something, and when they simply want attention. Intervening for situations where they child wants attention, may not eliminate its use when the child wants to avoid going to see the doctor.

**Fifth**, intervention involves changing social systems, not simply individuals. In considering intervention strategies, there are approaches that are directly applied to the child, those that are indirectly applied by changing the behavior of other people in the child's environment (such as changing the behavior of a teacher or a parent), and those that modify the environment itself. A question that might be asked is whether the problem is that the child is communicating inappropriately, or whether the social system is not listening to the child. Very often the approach is to make the child change to meet the demands of the system, as opposed to looking at how systems might better meet the needs of children.

Finally, lifestyle change is the ultimate goal of intervention. Dealing and coping with our children's problem behaviors is enormously stressful, due to worry and a sense of helplessness. Our goal should be children and families who are able to meet their needs and find satisfaction with their lives. Here are some ideas to help you cope:

First, be glad your child is trying to communicate. You may not like the method, but at least there can be some connection.

Second, choose your battles wisely. Not every misbehavior has to be dealt with. Not every troubling behavior needs to be labeled as misbehavior. Go after those that cause your child the most difficulty.

Third, don't feel like you have to go it alone. In fact, everyone in your child's environment needs to be on the same page in terms of responding to the problem. So mobilize as much help as you can from those other people.

Finally, take care of yourself. You are not simply the parent of a child with CHARGE. You are a person in your own right, with your own needs and goals. If you never have a chance to work on your own goals, you will become less and less useful to your child.