

LIFE EXPECTANCY IN CHARGE

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Children with CHARGE have been shown to have a 70% survival rate to five years of age (from actuarial survival analysis).^{1,2} The death rate is the highest in the first year of life.

The highest mortality is seen with a combination of CHARGE features of bilateral posterior choral atresia with either congenital heart defects or tracheo-esophageal.¹ If all three of the above are present, they offer the greatest risk of mortality or morbidity to the infant.

Patients with CHARGE have a high postoperative mortality.³ The reasons for this are postulated as hidden structural abnormalities of the larynx and/or pharynx with associated motor incoordination. This results in difficulty in intubation and problems after extubation. It is therefore important that the surgeons and anesthesiologists be aware of how complex these children are. When contemplating surgery in these children an experienced pediatric anesthesiologist should be involved, even if the surgery is rated as a minor procedure. It is advised that, if possible, a number of procedures be conducted under one anesthetic, thus limiting the number of anesthetics. A recent CHARGE girl at our hospital (IWK Grace Health Center, Halifax, Nova Scotia) undertook seven operations at the same time under one anaesthetic so it can be done!

References:

1. Blake, K., *et al.*, 1998, *CHARGE Association: An Update and Review for the Primary Pediatrician*, *Clinical Pediatrics*, pp. 1-16
2. Blake, K., *et al.*, 1990, *Who's in CHARGE a multidisciplinary approach*, *Archives of Disease in Childhood*, February, vol 65, no. 2, pp. 217-223.
3. Blake, K., *et al.*, 1993, *CHARGE association looking at the future - the voice of a family support group*, 1993;19,395-409.

CHARGE SYNDROME MANAGEMENT MANUAL FOR PARENTS

Introduction to the Developmental Sections

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Every individual with CHARGE is unique. Each has his or her own unique collection of medical problems. Each individual has his or her own personality and learning style. On top of that, each individual is in a different family setting and different medical and educational environments. We would love to be able to provide every family with a complete set of resources tailored to their needs. Obviously we cannot do that. As a consequence, every family will find different parts of this Manual helpful to them. Sometimes we have presented the same or similar information in a couple of different formats. Look it all over and choose the parts that are helpful for you. Use the loose-leaf format to reorganize, add, and delete information that is appropriate for your situation.

GET SOME TABS. There is a lot of information here. It will be much easier for you to find what you are looking for if you break the information up in to sections with tab dividers.

A note on “**DeafBlindness**.” We recognize that not all individuals with CHARGE have both hearing loss and vision loss. Nevertheless, much of the information is labeled “deafblind” or aimed mostly at that population. We did this for two reasons. First, families dealing with individuals who DO have dual sensory loss will have the most difficulty with development and communication and will have the hardest time finding appropriate resources. Second, many of these resources are just plain good resources regardless of the title.

Please don't skip a section just because it is labeled “DeafBlind” – take a look and see if it has something to offer you as well.

MEDICAL INFLUENCES ON DEVELOPMENT IN CHARGE

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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.

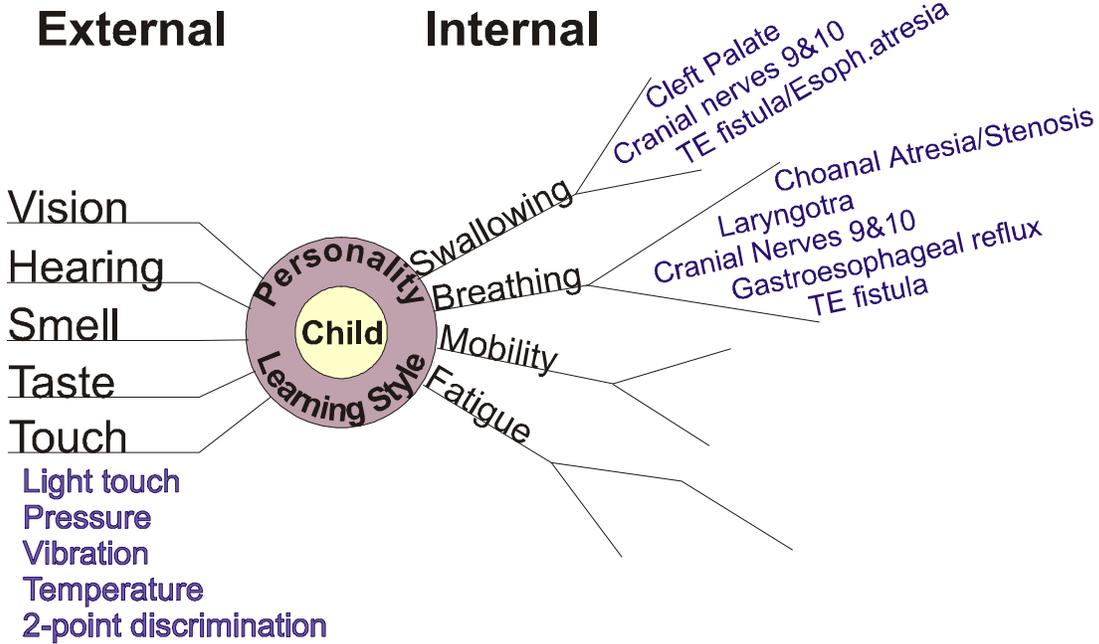
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CHARGE is such a complex disorder that many of the physical components have a profound effect on the child's development and understanding of the world. The above diagram breaks these influences down into those that are External and those that are 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.