

# GROWTH IN CHARGE: FOR THE PHYSICIAN

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Growth in children with CHARGE has special considerations in each of the three phases: Infant Phase, Childhood Phase, Pubertal Growth Spurt.

1. **Infant Phase:**

At birth, children with CHARGE Association usually have normal weights and lengths.<sup>1,2</sup> However, within the first 9 months of life there is a decline away from the normal growth curve, down to and often below the 3rd percentile. This pattern of growth may be related to repeat hospitalizations, poor feeding, major acute illnesses such as pneumonias and multiple surgeries. The infant phase of growth is mainly determined by nutrition and those children with CHARGE who maintain their weight in the early months are often the ones who have had major nutritional intervention, usually in the form of enteral feeding.<sup>1</sup>

2. **Childhood Phase:**

Although the growth rate is slower, the childhood phase of growth is the main determinant of final height as it lasts longer than the other phases. Adequate nutrition is important in this phase but also hormones play a role (thyroxine and growth hormone). Growth hormone deficiency is rare but there is an increased incidence in CHARGE.<sup>3,4</sup> Pre-adolescent children may have a more normal rate of growth although poor growth in infancy and lack of catch-up growth during childhood often result in a mean height at or below the 3rd percentile.

3. **Pubertal Growth Spurt:**

Growth deceleration associated with a delay in puberty occurs in more than 70 percent of children with CHARGE and this needs to be monitored closely. Growth in puberty involves the interaction of the sex steroids, especially testosterone and estrogen associated with growth hormone. There is anecdotal evidence that adolescents with CHARGE have fusion of their epiphyses at a later age and therefore often continue to grow in their early 20's.

The majority of children with CHARGE association have normal results on growth hormone stimulation testing. Arbitrary treatment with growth hormone in patients who have normal growth hormone levels has not been adequately studied and is probably not advised.

## **Nutrition    “Growth and Nutrition are Closely Related”**

Children need an adequate quantity and balance of food for optimal growth and development. CHARGE infants who have excessive surgery and a decrease in nutrition intake may require catch-up growth and their energy requirements may be 150 to 200 kcal/kg/d.

<b>Reference Values for Energy and Protein Requirements</b>		
<b>Age</b>	<b>Energy kcal/kg/d</b>	<b>Protein gm/kg/d</b>
0-6 months	115	2.2
6 mts.-6 yrs.	95	1.8
7 yrs-10 yrs.	75	1.2
11-16 yrs.	60	1

## **FEEDING ISSUES IN INFANCY AND EARLY CHILDHOOD**

### **Symptoms and Behaviors of Swallowing Problems (Also see Swallowing Section):**

1. The feeding history may include:  
Cough and choking, nasopharyngeal reflux, nasal congestion, food suctioned from the nose or from the tracheostomy.
2. Chronic chest difficulties.  
Recurrent pneumonias, apneas, frequent upper respiratory tract infections.
3. Other behaviors.  
The infant may have stresses on feeding such as sweating, gasping, straining, prolong or shortened sucking pattern. In children, other symptoms may be exhibited i.e. build-up of secretion, spiking temperatures, food remaining in the mouth, choking, eyes tearing or lurching the head forward and facial stress.

More than 90 percent of children with CHARGE have difficulty swallowing foods of different textures. As a consequence parents often discontinue trying to feed their children solid foods.

Children with bilateral posterior choanal atresia often have the most severe feeding problems and may manifest significant failure to thrive.<sup>4</sup> In some instances, there may be obvious physiological reasons for these problems, i.e. cleft lip and palate, facial palsy, choanal atresia, tracheo-esophageal fistula. Occult and hidden anatomical abnormalities of the pharynx and larynx have also been described.<sup>4</sup> These need to be considered in children and infants with feeding problems. One of the most difficult chronic management problems that families have to face is gastro esophageal reflux. There is no quick fix to the problem and different centers offer their expert team.

When gastrostomy tubes are used, children run the risk of oral pharyngeal hypersensitization, meaning that they reject substances and objects in their mouths, especially feeding devices. It is therefore important to try and keep stimulating the oral pharynx even though the child is being fed by gastrostomy or jejunostomy tube. Even with aggressive therapy many children with CHARGE continue to have feeding problems into pre-adolescent and beyond. Parents also report excessive abdominal colic similar of the type commonly found in infancy, which may present to school age children and beyond. The etiology of this colic is unknown but is probably organic in nature.

**General Management in Feeding a CHARGE Infant**

- Positioning of infant or child
- Use of different textures to find the ones that work
- Aspiration precautions
- Aggressive management during colds
- All of the above can be guided with a feeding team which should include an occupational therapist, physiotherapist, speech therapist, behavioral psychologist, and dietician.

**Breast Feeding** - often difficult in CHARGE infants because of their many surgeries but not impossible especially if expressed breast milk is used.

**Weaning:**

Solid foods are usually introduced between the ages of 3 and 6 months. After 6 months of age, milk only can lead to anemia and deficiencies in vitamins and iron. Lumpy foods, which are usually introduced at about 6 months, are hard for CHARGE children to manage. Different textures and tastes may also be a problem. Many families use pureed foods until childhood.

**Failure to Thrive:**

All children with chronic illness can have failure to thrive. Congenital heart disease or respiratory disorders alone can be responsible for failure to thrive. In CHARGE children, malnutrition may result from a combination of decreased intake, malabsorption and increased requirements because of increased work of breathing. Malnutrition from many of the aspects of CHARGE association can respond to enteral feeding.

Malnutrition must be recognized and accurately defined so decisions can be made about feeding. Evaluations are divided into assessment of past and present dietary intake, measurements and lab assessments as in the table above. The consequence of malnutrition is a multi-system disorder. Malnutrition worsens the outcome of illness, respiratory muscle dysfunction and may delay a child being weaned from a mechanical ventilator. Malnourished children are less active, less exploratory and more apathetic. These behavioural abnormalities are rapidly reversed with proper feeding.

<b>Assessment of Nutritional Status</b>
<b>Anthropometry</b> <ul style="list-style-type: none"><li>· weight</li><li>· height</li><li>· mid arm circumference</li><li>· skin fold fitness</li></ul>
<b>Lab Data</b> <ul style="list-style-type: none"><li>· low plasma albumin</li><li>· low concentration of specific minerals and vitamins</li></ul>
<b>Food Intake</b> <ul style="list-style-type: none"><li>· dietary recall</li><li>· dietary diary</li></ul>
<b>Immunodeficiency</b> <ul style="list-style-type: none"><li>· low lymphocyte site count</li><li>· impaired cell mediated immunity</li></ul>

### **Short Stature or Failure to Thrive:**

Short stature and failure to thrive are usually defined as height or weight below the 2nd or 3rd percentile (approximately 2 standard deviations below the mean). Serial measurements are the most helpful as they allow detection of change in the rate of growth. Growth failure can be defined from the child's growth parameters falling across percentile lines plotted on a growth chart. In this way, growth failure may be identified even though the child's height and weight are still above the second or third percentile. As most children with CHARGE have a normal birth weight and length, then it is often obvious within the first 9 months when they fall away from the middle percentiles. CHARGE children who suffer from malnutrition and chronic illness are usually moderately short but significantly underweight. In contrast, if growth hormone deficiency is the cause for growth delay, children are usually somewhat overweight for their height.

Short stature may cause psychological problems. Adults assume that the children are younger than their true age and so treat them inappropriately.

### **Growth Hormone**

Diagnosis of growth hormone deficiency is difficult because of the pulsator nature of growth hormone secretion. In pituitary provocation tests, a variety of stimuli are used to provoke growth hormone release. The most common is clonidine, glucogone and insulin induced hypoglycemia. The hypoglycemia induced by insulin is potentially dangerous. To avoid these complications, the test should only be performed in specialist centres. Growth hormone deficiency is treated with biosynthetic growth hormone, which is given by subcutaneous injection usually daily. There are other potential applications of growth hormone therapy, which is under study including children with different syndromes and those with chronic illness. CHARGE would fall within this realm, however, growth hormone is not being used at present for children who have not been diagnosed with growth hormone deficiency.

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## FEEDING AND GROWTH IN CHARGE: PARENT INFORMATION

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Although some children with CHARGE have growth hormone deficiency, much of the slow growth in CHARGE is likely due to nutritional status and general medical problems.

Difficulty in feeding is one of the most common and prolonged problems in CHARGE. As a consequence of poor feeding, the long term poor nutrition results in growth failure and can contribute to the developmental delay. Assessment of growth is important and below are the growth parameters that should be measured routinely in your children (3-4 times per year):

1. Weight - If possible, measurements should be on the same weight scales.
2. Length or Height - In children under 2 years old, length is measured lying horizontally. Because of balance problems and delayed mobility, children who are not comfortable standing to be measured should also be measured horizontally.
3. Head circumferences should be measured in the first 2 years of life. This is the widest circumference around the head called the occipital frontal circumference and is an indication of brain growth and overall size.
4. The measurements of growth parameters should be plotted on percentile charts. It is wise to keep a copy in your binder of your own child's growth progression and ask to be shown how to plot and read the charts.
5. Clinic - It is important to attend a growth and/or endocrine clinic. If growth hormone deficiency is suspected, your doctor may order special testing.

### **Feeding and Swallowing In Children With CHARGE (Also see Swallowing Section):**

Feeding problems are frequent and represent a permanent concern for most parents. Feeding studies may show that the infant can suck normally but liquid pools in the back of the throat without passing smoothly into the esophagus. Swallowing can be uncoordinated and also lead to gastroesophageal reflux. Children are often uncomfortable, show signs of restlessness when they are being fed, they may gag and aspirate (the food or liquid goes down into the lungs). These children have a hard time feeding orally. Some can handle pureed foods but cannot tolerate liquids or solids. It is important to find the consistency and type of food that your child will tolerate. Positioning and behavior modification when feeding are also important. A team of therapists should be involved with your child who has feeding problems. A therapy team includes a speech-language pathologist, an occupational therapist, a psychologist and a physician with some interest in feeding problems. If there is a center where there is a feeding team, this center should be consulted.

### **ANATOMICAL ISSUES WHICH HAVE AN IMPACT ON SWALLOWING:**

- ◆ Retrognathia - posteriorly placed mandible
- ◆ Micrognathia - underdeveloped mandible (jaw bone)
- ◆ Tracheoesophageal fistula (TEF) - presence of a fistula (a joining) between the tracheal and esophageal walls.
- ◆ Cleft lip and Palate - refers to a "split" or "separation" in the lip and/or palate
- ◆ Facial palsy - usually on one side but can be both sides. The side of the paralysis is the side where the eye has incomplete closure - more noticeable when the infant is crying.
- ◆ Choanal Atresia or stenosis

- ◆ Lower Cranial Nerves Involved in Swallowing and Sucking (IX X XI) - affects sensory (feel) and motor function (activity) involving sucking and swallowing.

#### **TESTS USED TO EVALUATE SWALLOWING:**

##### ***Barium Swallow:***

Assessment of anatomical structure function  
Examines the esophagus pharynx, and larynx  
Uses large amounts of liquid purees, and solids  
Positions the patient in the supine (lying down)  
Follows the bolus (watch as food goes down)

##### ***Videofluoroscopic Swallow Study (VFSS):***

Assessment of swallowing  
Examines the oral cavity  
Uses small amounts of liquid  
Positions patient in the upright position  
Does not follow the bolus

#### **ALTERNATIVE TYPES OF FEEDING:**

**Nasogastric Tube** - For short periods and for supplementation. This may not be a safe method for a child with a compromised swallow because of increased secretions produced by nasogastric tube, which can increase risk of aspiration.

**Gastrostomy Tube or Button** - Gastrostomy tube or button is a preferred choice for long-term supplementation if stomach function is intact.

**Jejunostomy Tube** - Preferred choice if severe gastroesophageal reflux is present.

#### Effects of Tube Feeding

1. Hypersensitivity of the oral cavity - as the mouth is not being stimulated, child gets sensitive to anything that is put in the mouth. Desensitizing the oral cavity is important. Always encouraging textures, tastes and sensitizing procedures even if your child is not feeding orally. (Seek the expertise of an occupational or speech therapist.)
2. After tube feeding is started even for a short period of time it is difficult to get the child back on to oral feeds.
3. Lumps and different textures may remain a problem into later childhood.
4. The tube may not work, dislodge, and need replacing. Granulation tissue (healthy scar tissue) can collect around the tube and so can infection.
5. Parents like the button as it gives their child better mobility and freedom to move.