



Factsheet 13

Eating and drinking issues in CHARGE syndrome

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Eating and drinking is a fundamental physical and social need. As social beings, we place huge significance on who we eat with and where. April Winstock says:

"For most of us, food and drink are sources of pleasure; they satisfy hunger and thirst, help to structure our day and provide opportunities for social interaction" (2007 p. 1).

If someone has a difficulty with eating and drinking, mealtimes can quickly become very stressful for all concerned.

People with eating and drinking difficulties may have:

- some sort of oral-motor problem
- physical and structural difficulties (such as a cleft or palsy)
- sensory-based eating or drinking difficulties
- difficulties organising the transfer of food to their mouth.

These can lead to a risk of:

- dehydration and malnutrition
- food going down the 'wrong way' into the airway (aspiration)
- choking
- infection, and
- faltered development of eating and drinking skills.

It is widely reported that children with CHARGE face significant challenges with eating and drinking – and these are often multi-factorial. They can be a combination of the difficulties described above with strong elements of both motor and sensory challenges.

A number of studies suggest that 79–90% of children with CHARGE have 'feeding difficulties' (Blake *et al.* 1998; Harvey *et al.* 1991; White *et al.* 2005 (cited in



Sanlaville and Verloes, 2007); Hartshorne *et al.* 2005; Dobbelsteyn, *et al.* 2008). In a recent study of CHARGE in the UK by Deuce *et al.* (2012), 84% of the UK cohort identified eating and drinking difficulties.

The challenges of eating and drinking for children with CHARGE are complex and, I believe, rooted in the Cranial Nerve dysfunction which underpins some of the characteristics presented by the syndrome. The diagram on page 4 shows how some of these relationships impact upon each other and demonstrates the complexity of the challenges that can affect eating and drinking development in children with CHARGE syndrome.



Characteristics of CHARGE associated with the motor, physical and sensory aspects of eating and drinking difficulties:

Characteristic	Motor/	Impact
Cranial nerve dysfunction	(Cr N I) Smell	Sensory The lack of smell impacting on developing taste system and appeal of food. Smell also plays a role in infant/ mother bonding and developing eating and drinking behaviours. Difficulties with the sense of smell are reported as occurring in >90% of individuals with CHARGE by Hartshorne <i>et al.</i> (2011).
	(Cr N V VII XII) Weak suck/ poor chew	Motor Physical Sensory This affects sensory and motor control of muscles and oral anatomy which leads to chewing and swallowing difficulties. Dobbelsteyn <i>et al.</i> (2008) suggest 77% of children with CHARGE are affected.
	(Cr N V IX X XI) Swallowing difficulty	Motor Physical Sensory Affects sensory and motor control of muscles and oral anatomy required for chewing and swallowing. According to Hartshorne <i>et al.</i> (2011) 70–90% of individuals with CHARGE are expected to have swallowing difficulties with aspiration.
	(Cr N X) Gastro-Oesophageal Reflux (GOR)	Motor Sensory This is when the liquid content of the stomach regurgitates (backs up or refluxes) into the oesophagus. Between 50–89% of individuals with CHARGE are affected by GOR (Dobbelsteyn <i>et al.</i> 2005; Salaville and Verloes 2007). A study by Deuce <i>et al.</i> (2012) found 45% of children were medicated for reflux in the UK.
Respiratory problems	Motor Physical	Respiration may be interrupted during eating and drinking. A Tracheotomy may interrupt the suck-swallow-breath synchrony. Other anomalies found in children with CHARGE are: Laryngomalacia (floppy/collapsing larynx) Tracheomalacia (floppy/collapsing airway) Subglottic stenosis (narrowing of subglottic airway).
Tracheosophageal fistula (TOF)	Physical	A TOF precludes a normal swallow and according to Hartshorne <i>et al.</i> (2011) occurs in 15–20% of children with CHARGE.
Cleft	Physical Motor	A cleft of the lip or palate prevents negative pressure which impacts on the ability to suck. Hartshorne <i>et al.</i> (2001) reported this condition to be found in 15–20% of individuals with CHARGE.
Choanal atresia/stenosis	Physical Motor	Hartshorne <i>et al.</i> (2011) reported the occurrence of this anomaly in 50–60% of individuals with CHARGE. This can be either a partial or complete blockage that impacts on breathing and the suck-swallow-breathe synchrony. <i>(chart continues on next page)</i>



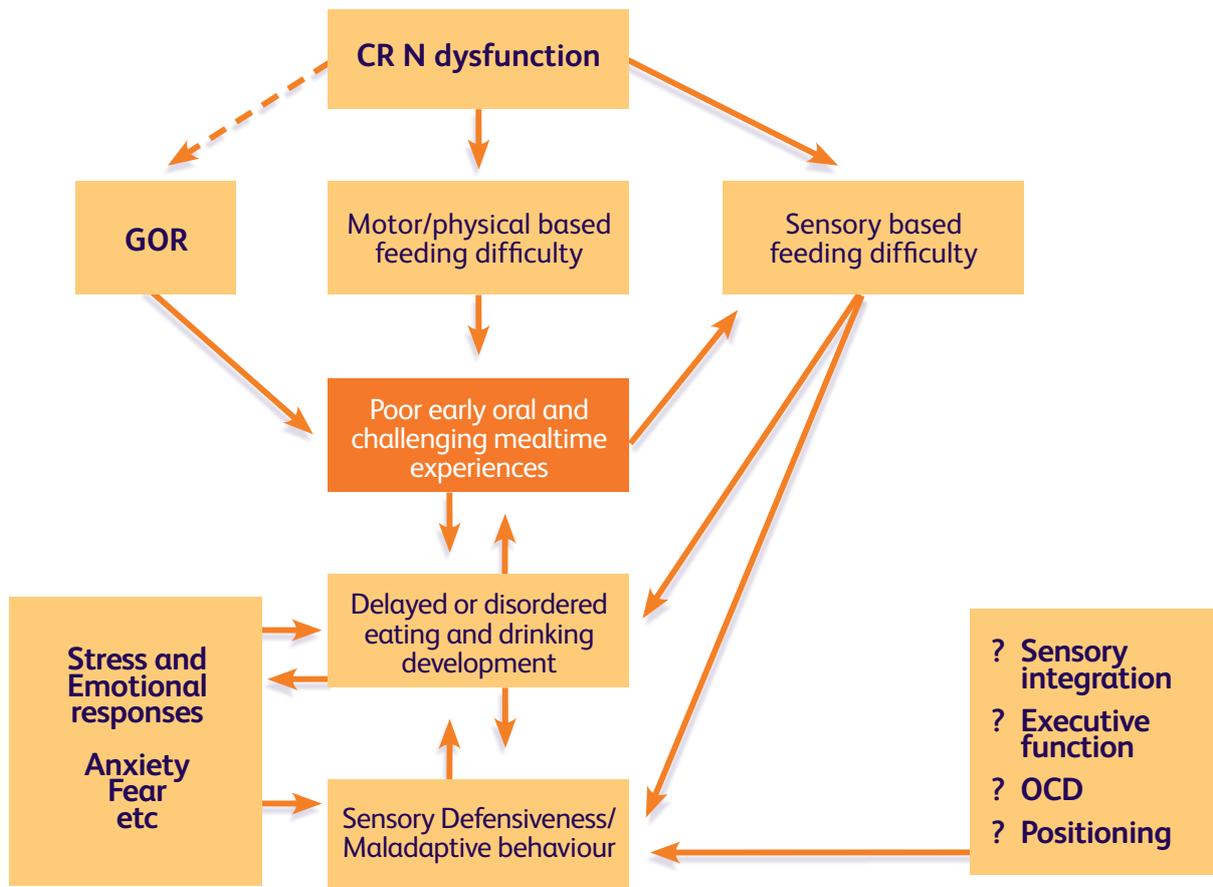
(Continuation of chart from previous page)

Characteristic	Motor/	Impact
NG tube feeding	Sensory	<p>There is no specific data, but anecdotal evidence suggests that up to 92% of children with CHARGE have a history of tube feeding.</p> <p>The potential impact of these could be:</p> <ul style="list-style-type: none">• reduced early oral experiences, reducing development of the link between satiation and oral experience.• a delay in starting oral feeding can result in missing key critical periods for development. <p>It can also adversely affect:</p> <ul style="list-style-type: none">• regulation of state which develops during the first 2–3 months.• the development of attachment (2–6 months) where mealtimes become a social time.• the development of separation and individualisation – sense of control, sense of self and differentiation of emotional and bodily states (6–36 months).
Numerous medical interventions	Sensory	<p>Again there is no specific data, but the primary need in a child's early days is to establish medical stability – and this may involve numerous interventions.</p>
Sensory integration dysfunction	Sensory	<p>The child is unable to maintain a level of alertness or arousal so that they are unable to sit still to eat and drink, or fall asleep during eating and drinking.</p>
Executive function difficulties	Other	<p>This creates difficulties in starting mealtimes and initiating eating and drinking. A child may even find it difficult to stop eating despite being full.</p>
Obsessive compulsive disorders	Other	<p>Difficulty starting mealtimes without a specific routine or taking control of specific aspects of the mealtime.</p>

There are clear relationships between motor/physical and sensory-based feeding difficulties. It is also common for motor-based feeding difficulties to be exacerbated by sensory-based experiences with the development of maladaptive behaviours (such as: food refusal, gagging, breath holding, food holding in or expulsion from mouth).



THE RELATIONSHIPS BETWEEN DIFFERENT FACTORS AFFECTING EATING AND DRINKING DEVELOPMENT



Key points:

- Poor early oral feeding and/or challenging mealtime experiences can lead to the development of sensory feeding difficulties, and delay eating and drinking skill development.
- GOR complicates things further, exacerbating negative experiences and associations with mealtimes.
- Sensory integration, executive function difficulties, Obsessive Compulsive Disorder and suitable positioning can have an additional impact.
- Other factors that can compound eating and drinking difficulties include: stress/emotion, the development of sensory defensiveness and maladaptive behaviours.
- Mealtimes are made more challenging by the true multi-sensory impairment experienced by children with CHARGE, particularly in relation to the unique challenges of sensory integration and executive function difficulties, and OCD.

Approaches to try

- Messy play is a great tool that can be used to help a child to explore food in a fun and relaxed way. Playing with food can be a good way to help a child overcome any fears that may have developed for a variety of reasons.

A structured approach to exploring textures – from those which fall away and break contact with the child’s fingers easily, to those that require repeated wiping and stick to the hands – can be a helpful approach to building up tolerance. When exploring any textures or in messy play activities a hand-under-hand exploration technique can be a safe way for the child to develop trust and experience new textures.
- A proactive management approach which promotes positive mealtime experiences and reduces the chance of sensory selectiveness developing. Once safety and nutrition have been safely managed, it is also important to focus on the oral-motor development. These sorts of programmes should be put together by Speech and Language Therapy/



Occupational Therapy colleagues with the appropriate skills and training.

- Management should be multi-professional with practitioners adopting a strong sense of trans-disciplinary working.
- Transitions should be planned and reviewed frequently. Practitioners have a role to promote transition to oral feeding whilst managing a balance between progress and safety of swallow, nutritional status and emotional well-being.

Many children with CHARGE make the transition eating orally, so we need to make sure that we minimise the duration of non-oral feeding and facilitate the transition to oral eating.

- Investigations should be undertaken for GOR and aspiration for all individuals with CHARGE syndrome to exclude or take appropriate action to alleviate implications of GOR/aspiration.

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GLOSSARY

Choanal stenosis/atresia: a narrowing or blockage of the passageway between the nose and the pharynx by tissue.

Executive function: the cognitive process that regulates an individual's ability to organise thoughts and activities, prioritise tasks, manage time efficiently, and make decisions.

Hand under hand: an approach used with children who are multi-sensory impaired to support mutual exploration and communication. The adult places their hands under the child's to guide and support them to engage with their environment.

Tracheoesophageal fistula (T-E fistula or TOF): an abnormal connection (fistula) between the oesophagus and the trachea.

Sensory integration: the neurological process that organises sensation from one's own body and the environment – which makes it possible to use the body effectively within the environment.

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