**Presenter Information:**

**Steve Rose** is a Health Professions Council registered speech and language therapist specialising in working with people with deafblindness. He has previously worked as an intervenor and for Sense working with family support groups until returning to study to train as an SLT. Graduating from UCL in 2003 he subsequently worked in special schools in North London with children with physical disabilities, sensory impairments, autistic spectrum disorders and learning difficulties. Steve has particular interest in the development of eating and drinking skills and early interventions, including parent-child interaction therapy. He has recently completed his MEd in deafblindness at Birmingham University and currently heads Sense Children’s Specialist Services.

**Presentation Abstract:**

It is well documented that individuals with CHARGE syndrome may face difficulties with eating and drinking. There is emerging literature that documents the prevalence of these issues and support intervention for such difficulties.

This presentation explores literature of the prevalence, nature and range of difficulties and the subsequent challenges presented. The features of CHARGE that relate to difficulties with mealtimes are also identified. It will outline some observations on the nature of the challenges facing children with CHARGE seen at our centre in North London. These challenges will be discussed in the context of case studies to illustrate individual challenges and possible management strategies.
It is recognised that the early oral experiences of children who experience challenges eating and drinking have an impact on later development and prognosis. The involvement of cranial nerve dysfunction in CHARGE leads to a high percentage of children having eating, drinking and swallowing disorders – up to an estimate of 80% of all cases (Sanlaville and Verloes 2007). In addition there are numerous other factors and co-occurring features that impact on these difficulties, combined these present significant challenges for the child that are often seen as ‘behavioural feeding issues’.

Case studies will identify some of the ‘mealtime behaviour’ of children seen at our centre and begin to develop some understanding of the challenges facing this group of children and the sort of support that could help to overcome them. This is an area where there is less specific literature and it is hoped that this paper will begin to contribute to a greater understanding of these issues for this group of children.

3rd Professional Day & 11th International CHARGE Syndrome Conference
The challenge of mealtimes for children with CHARGE syndrome

Steve Rose
Speech and Language Therapist / Head of Children’s Specialist Services
11th International CHARGE Syndrome Conference
July 25-28 2013

The plan for this session is to

• explore the nature of eating and drinking difficulties in its broadest context.
• look at the neurological control required to eat and drink, including to trigger the swallow (a pattern elicited response).
• look at which aspects of CHARGE impact on eating and drinking, exploring themes which have emerged from the evidence base in this area.
• identify other issues based on observations made within our service paying particular attention to sensory-based feeding disorders illustrated with case studies.

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

(Winstock 2007 p.1)

Challenges

1) children who present with some sort of oral / motor problem (dysphagia)
2) children who present with a sensory based feeding difficulty
3) children who have difficulty organising the transfer of food to their mouth.

“The primary goal in managing eating and drinking difficulties should address the child’s safety [of swallow], nutrition and hydration needs along with emotional well-being of the whole family.”

(Winstock 2007 p.128)
Risks

Dysphagia:
- Dehydration and malnutrition (leading to organic failure to thrive)
- Aspiration
- Choking
- Faltered development of eating and drinking skills
- Infection (secondary to previous risks)

Sensory based feeding difficulties:
- Dehydration and malnutrition (leading to non-organic failure to thrive)
- Faltered development of eating and drinking skills

Neurology and swallowing

The normal swallow is a four stage process
- Oral preparatory stage – food is manipulated in the mouth.
- Oral stage – tongue pushes food back.
- Pharyngeal stage – swallow reflex triggered, food moves down through pharynx.
- Oesophageal stage – food moves down to the stomach.

Cranial Nerves involved in the Innervation of the oral and pharyngeal muscles and structures.

I     Olfactory Nerve
V    Trigeminal Nerve
VII  Facial Nerve
IX   Glossopharyngeal Nerve
X    Vagus Nerve
XI   Spinal Accessory Nerve
XII  Hypoglossal Nerve

CHARGE

79 - 90% of children with CHARGE have feeding difficulties


Characteristics of CHARGE associated with eating and drinking difficulty.

Motor related
- Cranial Nerve dysfunction
  - Smell (Cr N I)
  - Weak suck / poor chew (Cr N V VII XII)
  - Swallowing difficulty (Cr N V IX X XI)
  - GOR (Cr N X)
- Respiratory problems
- Cleft (lip, palate or both)
- Choanal Atresia

Sensory related
- Cranial Nerve dysfunction
- GOR
- Long term NG tube
- Discomfort (related to GOR?)
- Numerous medical interventions

Motor based feeding difficulty
Sensory based feeding difficulty
GOR
Motor based feeding difficulty
Sensory based feeding difficulty
Post early oral and challenging mealtime experiences
Delayed or disordered eating and drinking development
Sensory Defensiveness/ Maladaptive behaviour
Stress & Emotional responses
Anxiety, Fear etc

? Sensory integration
? Executive function
? OCD
? Position
Our Assessment Centre

A: Children who are being non-orally fed
B: Children who are managing to eat orally
C: Children who are coping with oral eating (but it’s not all plain sailing)
D: Children who are coping with oral eating and experiencing delay with the transition to a normal diet

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Case studies

Preparing for mealtimes

• Take time
• Involve the child in preparation
• Set up the environment
• Sit in a good seating position
• Prepare the face / hands (massage?)
• Use the right cues (more, enough, stop!)
• Establish sensory equilibrium – Sensory diet activities before the mealtime
• Think about starting, continuing and finishing
• Prepare the right texture foods (and separate tastes)

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During mealtimes

• Use the right utensils
• Relax!
• Don’t have a battle
• Take a break – consider little and often?
• Think about the social elements
• Consider use of distractors and motivators

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Food Play Texture Hierarchy

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Conclusions for management

- Proactive management promoting positive mealtime experiences
- Whilst anatomical and medical aspects are being managed the introduction of a positive oral stimulation program is considered and where possible implemented
- Mealtime assessment should be undertaken between SLT and OT colleagues and within the context of a sensory integration approach.

"Need for sensory integrative approach to management of eating and drinking. Feeding is a highly integrated multisystem skill and many contemporary clinicians support the approach that it is not helpful to focus solely on the child's mouth but to consider the whole child, including the environment, the child's sensori-motor profile and the context of mealtimes when developing a treatment plan" (Maune 2007 p.4)

Conclusions for management

- Multi-professional, trans-disciplinary management
- Transitions are planned and reviewed frequently, with a duty to promote transition to oral feeding.
- Investigations should be undertaken for GOR and aspiration for all individuals with CHARGE syndrome
- Intervention is ongoing, multidisciplinary and multi-sensory system

"All children with CHARGE syndrome to be followed proactively by a multidisciplinary team consisting of gastroenterologist, ENT surgeon, nutritionist and other feeding specialists (occupational therapist and/or speech and language pathologist) to ensure appropriate management of feeding issues"

Dobbelsteyn et al 2008 p 132

"The presence of gastro-oesophageal reflux and aspiration early in life will increase the probability of ongoing feeding problems..."

Dobbelsteyn et al 2008 p 132
“follow up should involve multi
disciplinary feeding team and
parents should be made aware
of the potential long term
feeding issues”
Dobbelsteyn et al 2005 p. 99

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