DYSPHAGIA AND NUTRITIONAL ISSUES IN NEURODEVELOPMENTAL DISORDERS
CHALLENGES AND DILEMMAS
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Multidisciplinary Clinical Services
(Paediatric & Adult)
• Current population - >800 children and adults with severe neurological impairment (80% CP) and complex health needs
• Referral sources
  • Clinics
    – Dysphagia/Nutrition, Bone Health, Respiratory, Complex Epilepsy/ID, Tuberous Sclerosis
    – Down Syndrome, Rett’s Syndrome, General Disability
• Clinic personnel Paediatric
  – Gastroenterologist, Developmental Paediatrician
  – Speech Pathologist, Dietitian and Students, CNC
• Clinic Personnel Adult
  – Disability Physician, Speech Pathologist, Dietitian, Dentist, CNC, Students
  – Access to Surgeon, Nutrition Support Team

Paediatric Feeding Clinic
• Feeding Difficulty:
  • Often Post neonatal (rocky course) – prolonged hospitalisation
  • Behavioural – e.g. ASD

Management Challenges
• Complex medical and psychosocial
• Communication (multicultural)
• Consent Issues
• Ix – complex, may need G.A.
• Declining health
• End of life discussion

Cerebral Palsy
Associated conditions/problems
• Motor (80%) spasticity
• Sensory – tactile/vision/hearing
• Nutritional
• Cognition
• Communication – 50% need assistive devices
• Epilepsy ~ 45-50%
• GIT/dysphagia/nutrition
• Bones/orthopaedic – hips, spine, pain, musclesd
• Respiratory

Autism Spectrum Disorder
• Heightened sense of smell is common & can impact on acceptance of foods
• Many children have heightened sensitivity to changes in food texture
• Heightened sensitivity can cause gagging
• Motor planning difficulties can affect oral motor skills development & self feeding
• Communication difficulties can affect parents’ ability to read signals of overload or pleasure surrounding eating
Assessment

- Nutritional status and swallowing ability
- Reflux & upper GI disease
- Bowel Management
- Chest disease
- Bone health
- Other co-morbidities e.g. epilepsy, mental health

Dysphagia Assessment

- Observation in familiar surroundings first and position well
- Experienced Speech Pathologist
- Variety of consistencies

Investigations for dysphagia

- Good History and exam
- Endoscopy
- Video-fluoroscopy (MBS)
- Nutritional parameters
- Helical CT chest
- Bone Density Study
- Manometry
- Limited use – Ba study (delayed emptying)

Nutrition Assessment

- History + food records
- Anthropometry
- Skin folds
- DEXA
  - Body fat
  - Lean mass
- Eyeball test

Nutritional Intervention

- Assess diet & increase calories (Dietitian)
- Alter consistency of food and swallow (Speech path)
- Positioning (Physio)
- Dental consult

GORD

- Blood tests e.g. iron stores
- Endoscopy & biopsy
- pH study
- Barium study - possibly
- Nuclear medicine milk scan
Overall Management Goals

- Adequate nutrition
- Treatment of GOR constipation etc
- Stable respiratory function
- Developmentally appropriate oral sensory and motor function
- Fostering a positive feeding relationship
- Participation in the social & communication activities during mealtime
- Oral feeding (not always)

Handling sensory challenges of mealtimes

- Make changes slowly
  - Start with a familiar food and add very small amounts of a new one
- Mask the smell of new foods
  - Lids on cups or straws are often helpful, use chilled foods
- Provide activities that reduce anxiety and increase focus of attention before meals
  - Deep touch, vestibular stimulation, calming music

Medical Management

- What is ideal nutrition?
- Maximise nutrition
- Treat GOR/gastritis
  - PPI, Triple therapy – H. Pylori
  - Steroids for EEO
- Altered textures & thickened fluids
- Bowel Management Plan
- Dental hygiene plan
- Chest management plan
- Address bone health

Indications for non-oral feeding

- Weight and nutrition
  - Poor oral intake, weight gain and growth
  - Low micronutrients – Iron, Calcium, Vit D
- Initial Interventions
  - High energy additives – fats, cream
  - Supplementary drinks, formulas or puddings
  - NGT
    - Consider safety, support, practicalities

Indications for NGT feeding

- Short-term
  - Post surgical procedure
  - Post illness
  - Increase in seizures (alertness, aspiration)
  - Safety issues – oral aversion and mucosal damage
- Move to gastrostomy soon for long term management
  - Dysphagia (Unsafe swallow)
  - Poor chest health (Recurrent pneumonia/chest infections)
  - Inadequate Nutrition

Indications for Gastrostomy

- Inadequate nutrition
- Aspiration/ choking
- Recurrent lung infections
- “Top-up” calories, fluids + medication administration during prolonged periods of altered consciousness
- NB:- Surgical gastrostomy placement (because of risk of perforation with PEG)
Decisions making around gastrostomies

- Reality
  - Reality
  - Longer surgical waiting lists
  - May take up to 2 years to gain consent
  - Issues around no longer being able to eat orally
  - Tube/button lasts variable length of time 3mths to 2 years
- Which device?
  - Majority of children have mick-key low-profile devices placed
  - Adults usually have gastrostomy tube initially, and then convert to button
  - Advantage – change at home
  - Disadvantage – easily burst, easily removed, may become blocked if not flushed adequately i.e. after medication.

Indications for Fundoplication

- Ongoing GOR disease
- Recurrent chest infections due to reflux

Inadequate Nutrition despite Gastrostomy

- Failure to use gastrostomy
- Incorrect feed regime and/or financial issues
- Gastric stasis, delayed emptying
- Chronic infection
- Next step – jejunostomy or gastro jejunostomy
- TPN considered if all else fails

Drugs & Gastrostomies

- Use local pharmacist
- Gastric irritation – Valproate should be given with feeds
- Poor absorption – Phenytoin should be given 2 hours prior or after feeds
- Enteric coated tablets must never be crushed.
- Omeprazole should not be crushed – break open and mix with water, or request in syrup form.
- When possible medications should be in liquid form to prevent tube blockages.

Site management

- Daily clean with soap & water
- Reduce leakage and keep dry
- Steroid creams for rashes
- Barrier creams to protect skin – cornflour, (padding not advised)
- Monitor hyper-granulation – hypertonic saline
- Monitor infection
- Check if secure/rotate daily

Issues around Treatment - Barriers

- Lack of reliable data/little literature
- Nurture & "normality" of oral feeding
- Cultural aspects of feeding
- Superstition
- "Control" / Independence of feeding
- "Size" of child/adult
- Resistance to ix / simple interventions
- Equipment/therapy/dietary costs
- Duty of Care
- Prolongation/Quality of Life
- Time/information/support
Quality of Life

- Sullivan et al., 2005 – Prospective study children with severe neurological impairment.
- Post gastrostomy placement – 6 and 12 month follow-up
- Improved weight, length, skin fold thickness
- Improved health
- Improved parental concern re health of child
- Decrease in chest infections
- No change in hospitalisations

Summary

- Complex conditions
- Simple interventions
- Multidisciplinary approach
- Access and availability
- Time/information/support/advocacy
- Quality of life focus

Bone health in Cerebral Palsy

- Determinants – bone health:
  - Calcium/Vitamin D/Nutrition
  - General health, body weight, medication
  - Genetics 80%
  - Exercise 17%
  - Gonadal hormone status
  - Lifestyle 20%

Management

- Maximise mobility (Walking/ Standing/ Vibration)
- Mineral homeostasis (Calcium, Vit D) – supplementation decreases rate of falls and fractures.
- Needs sunlight
- Some medications e.g. phenytoin increase metabolism of Vit D
- Pubertal progression
- Bisphosphonates
  - Fracture
  - Pain
  - Orthopaedic

Respiratory Disease

- Lung disease common in CP
- Multi-factorial
- Simple interventions/therapies
  - ↓ hospitalisations
  - ↑ QOL
- Sleep disordered breathing
  - Under-diagnosed
  - Under-treated

Respiratory Disease Contd…..

- Asthma is over diagnosed and under treated
- Asthma Symptoms due to :
  - Associated GOR +/- aspiration
  - Poor mucociliary clearance
  - Upper airway obstruction
  - Chronic lung disease
### Chronic Lung Disease
- Aspiration with swallowing  
  - Stop oral feeds  
- Reflux and aspiration  
  - Antireflux surgery  
- Salivary aspiration  
  - Consider use of Botox  
- Recurrent infection in established bronchiectasis

### What do we do?
- History & examination (anatomy, upper and lower airways + GIT)  
- Ix – Sputum, CXR, Biochemistry, G.I. Ix e.g. Ba study  
- Assess aspiration with video-fluoroscopy (MBS)  
- Assess respiratory status with helical CT  
- Assess GOR with endoscopy  
- Assess sleep disorder with sleep study

### What do we do next?
- Refer to respiratory physician  
- Mealtime management plan (therapy team + dietitian)  
- Dental hygiene and saliva mgmt plans  
- Chest management plan  
  - Positioning (feeding +sleeping)  
  - Physiotherapy  
  - Immunisations  
  - Mgt of infections  
- Review of seizure mgmt

### What do “they” do?

#### Respiratory Ix
1. For acute/recurrent infection  
   - CXR +/- Helical CT Chest  
   - Sputum culture ± blood culture  
2. For aspiration  
   - Bronchoscopy (rarely)  
3. For sleep disturbance  
   - Sleep study

### End of Life
- Discuss end of life care plan – challenging  
- Patient, family, health professionals  
- Ethical, cultural, religious issues  
- Parents have ethical and legal rights except for decisions which cause harm – then illegal!

### Transition Paediatric to Adult Services
- Design not Default  
- Planning/Preparation  
- Patient stability  
- Comprehensive medical history  
- Summary for ED  
- Contact details – Specific Health Professionals  
- Case Manager – eg GP  
- Psychosocial issues  
- Explain differences especially numbers!
Paediatric –Adult differences

<table>
<thead>
<tr>
<th>Paediatric</th>
<th>Adult</th>
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</thead>
<tbody>
<tr>
<td>Paediatric case mgr</td>
<td>No case mgr</td>
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<tr>
<td>Variety of clinicians with ID expertise</td>
<td>Little exposure to ID</td>
</tr>
<tr>
<td>Consent by parent</td>
<td>Consent by ?</td>
</tr>
<tr>
<td>Multidisciplinary care</td>
<td>Subspecialist care</td>
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<tr>
<td>Lifelong records</td>
<td>Records often limited</td>
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<tr>
<td>Optimism</td>
<td>Realism</td>
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<tr>
<td>Long association with clinicians &amp; system</td>
<td>Trust takes time</td>
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<tr>
<td>Child and family centred</td>
<td>Seen as less “warm”</td>
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Transition (Cont)

• Comprehensive – clear medical summaries, including psychosocial and family history
  • Full seizure, medication and Ix history
    – incl failed medications and adverse effects
    – May redo investigations e.g. MRI, EEG, Telemetry
• Parent/carer expectations
• End of life – Palliative care, not withdrawal of care