Long-Term Management of the Child with Cerebral Palsy: Parents’ and Pediatricians’ Perspectives

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Definition of Cerebral Palsy

Cerebral palsy describes a group of disorders of the development of movement and posture that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, cognition, communication, perception and/or behaviour, and/or by a seizure disorder.

Cerebral Palsy

1. Symptom complex, rather than a specific disease
2. Umbrella term covering a group of non-progressive, but often changing motor impairment secondary to lesions or anomalies of brain arising in early stages of its development
3. Striated muscles = motor
Concerns of Parents: Top Ten

1. Eating
2. Rumination/vomiting
3. Constipation
4. Drooling
5. Development
   - Walking
   - Talking
   - Toileting
6. Sleep
7. Growth
   - Heavy
8. Puberty
   - Menstruation
   - Pregnancy
9. Sex
10. Transition/Future
Concerns of Pediatrician: Top Ten

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Concerns are Interconnected

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Disorders of Feeding and Swallowing

Implications of Development, Growth, Nutrition, Respiratory Health, Gastrointestinal Function, Parent-Child Interactions and Overall Family Life
Feeding Problems

I. Stallings

3 tertiary medical centers
A. N=154 - Diplegia or Hemiplegia
   30% - under nourished
   14% - overweight
   Children in the youngest group were most at risk for poor nutritional status

B. 86% children with quadriplegia - feeding
   37% children with diplegia or hemiplegia

II. North American Growth in CP Project

N = 230
Moderate to Severe CP
58% - feeding problems

III. Oxford Feeding Study

N = 440
frequent choking = 56%
stressful and prolonged feedings = 43%
vomiting = 22%
Considerations for Oral Feeding of Children with Cerebral Palsy

1. Dysphasia is more common in children with severe motor impairment
2. Aspiration is a common complication of dysphasia and is usually silent
3. Feeding history is important but often misleading
4. Feeding interruptions, duration of individual feedings, and consumed food textures are useful historical estimates of feeding efficiency
5. Observation of oral feedings is important
6. Weight gain is a good measure of oral feeding efficiency
7. Even though oral feedings may be difficult, they are important to children and families
8. Malnutrition usually presents in early infancy and is rarely resolved by continued oral feedings
9. Chronic lung disease is the most common sequela of aspiration
10. Gastroesophageal reflux is very common and can complicate oral feeding, appetite, growth, and respiratory status

Gastroesophageal Reflux

G-E sphincter is incompetent
- Allows stomach contents to reflux into the esophagus
- Common - spastic quadriplegia
- Acidity → inflammation in esophagus → pain → “heart burn” usually after feeding
- Posturing of neck - Sandiford syndrome
- Vagal nerve irritability caused by inflammatory process in esophagus (decrease heart rate).

Dx:
- pH probe
- UGI

Tx:
1. H2 Blocker (e.g. Ranitidine (Zantac), selectively antagonizes histamine H2 receptors)
2. Proton Pump (e.g. Omeprazole (Prilosec), inhibits gastric parietal cell hydrogen potassium ATPase)
3. Nissen Fundoplication
4. Gastrojejunal feeding
5. No good medication to increase emptying of the stomach (Cisapride, Metaclopramide)

Also assessing:
1. Growth
2. Rumination/Vomiting
3. Aspiration Issues
4. Chronic Lung Disease
Gastroesophageal Reflux

Comparing Fundoplication vs. GJ Feeding tubes

N=366
Fundoplication=323, GJ=43
No difference in aspiration pneumonia or mortality

Oral Aversion

Behavioral therapy works
  “Time to eat”
    Speech Pathologist
    Nutritionalist
    Social Work

Can d/c GT
Nutrition

CP imposes an extraordinary metabolic burden associated with spasticity and disordered movement

~ 50% - with significant under nutrition
**Nutrition**

Measure - Weight
1. Wheelchair scale

Measure - “Length”
1. Upper-arm length (UAL) - arthropometer
2. Tibial length (TR) - medial joint line of knee to inferior rim of medial malleolus
   - steel tape
   - easiest to measure
3. Knee height (KH) caliper
4. Skinfold thickness

Need to assess nutritional status
Weight-for-Height growth curve
10% bedridden
25% for children in wheelchair (decrease muscle mass)

Nutrition

Bone issues

1. Osteoporosis - decrease bone mass
2. Osteopenia - decrease bone mineral density

Poor bone formation - multifactorial
- Occurs primarily in non-ambulatory children
- Do not develop normal strength and size → unless a normal amount of stress is applied
- High rate of low energy fractures
- Child cried - ~24° before being seen
Growth and Health in Children with Moderate-to-Severe Cerebral Palsy

Growth patterns and relationship to health
6 sites, N = 273

Subjects with best growth had fewest days of health care usage (days in bed, days in hospital, visits to doctor or emergency Department) and fewest social participation missed (days missed at school or of usual activities for children and family).

Children with the worst growth had the most days of health care use and most days of participation missed.

Indications for Gastrostomy Tube

1. Dysphasia resulting in under nutrition
   Parent’s concern - heavy

2. Aspiration with associated respiratory disease

3. Insufficient fluid intake and/or refusal of oral medications

4. Excessive effort of stress during oral feeding

5. Mother - issue
Gastrostomy

Perioperative mortality rates low

Minor Complications
- leakage from stoma
- cellulitis
- excessive granulation tissue formation

Nissen Fundoplication
Drooling - Sialorrhea

10-40% in children with CP

Health

Cosmetic issues
Drooling

- Multiple therapeutic interventions
- Stepwise progression
  - Behavior therapy
  - Pharmacotherapy
  - Surgical procedure

Drooling

Treatment:
- Oral motor stimulation therapy
- Behavior modification
- Stylish scarves (bibs)
- Medications
- Botulinum-toxin injections
- Oral appliances
- Surgery
Drooling

Behavior modification:
For children who are aware enough to obey commands and cooperate with training
- Exercises to improve lip seal and tongue movement
- Trained Speech and Language Therapist
Drooling

Medications
- Anti Cholinergics (antagonizes acetylcholine receptors)
  Glycopyrolate (Robinul)
  Cogentin
  Scopalamine patch

Side effects:
- Constipation, dry mouth, blurred vision,
  urinary retention, nausea, weakness,
  tachycardia, confusion, irritability, behavioral changes
Drooling

Botulinum-toxin Injections

- Maybe effective inhibitor of salivary gland activity
- Maximum effect 2-8 weeks
- Ultrasound
Drooling

Palatal Training Appliances

↑ Oral Awareness
↑ Tongue movement and promote swallowing initiation
   > 6 years of age
   Dental Impression
   ? Work
Drooling

Surgery

- Parotid duct ligation/relocation/coagulation
- Removal of salivary gland - Submandibular Glands
- Submandibular Duct Translocation with sublingual gland excision

- Surgical complications
- Somewhat effective
Aspiration

1. Passive drainage of saliva (OPA = oropharyneal aspiration)

2. Dysphasia - food

3. GE Reflux - stomach contents

Present: Chronic cough
        Pneumonia
        Irritable
        Posturing
        No symptom - silent aspirator
Aspiration

Many of us aspirate small amount of gastric/oral secretions

Dx: videofluoroscopy - sensitivity and specificity
    ph probe (GE Reflux)

Chest x-ray = relative insensitive for detecting early or subtle signs of
damage to the airways or parenchyma

1. High resolution chest computer tomography (CT) document
   structural changes and detect early stages of bronchiectasis,
   bronchial wall thickening, minor atelectasis, patchy over-
   inflation

2. Ventilation Perfusion Scan - gives an estimate of function,
   helps determine whether structural damage on CT correlates
   with lost function
Aspiration

Limited evidence to support risk:
1. What affects the development of respiratory disease?
2. What is safe versus unsafe?

Not straightforward relationship between proven OPA and respiratory morbidity

Similar clinical situations can culminate in different pulmonary outcomes

Given lack of evidence-based guidelines - accepted pragmatic argument has been that any clinically demonstrated aspiratory may be dangerous

Prevention of Aspiration

Food

1. Postural management
2. Texture modification
3. NPO
   Parents = Failure
   Eating - fundamental right enjoyment (taste)

OPA

1. Pulmonary Vest Therapy
   - N=12
   - quadriplegia
   - poor cough
   ▼ Pneumonias
   ▼ Hospitalization
     More effective suctioning
   ▼ Seizure frequency

Constipation

1. Adverse consequences
   - Behavior Problems
   - Poor feeding - decreased appetite
   - Pain
   - Vomiting
   - Rectal bleeding
   - Social stigma/emotion
   - Emotional stress associated with incontinence
   - Megacolon
   - Bowel obstruction

2. What is problem?
   - Frequency - fewer than 3x/week
   - Consistency of stool - hard, large
   - Difficulty passing - blood, fissures, hemorrhoids, rectal prolapse, dermatitis, abscesses
   - Diarrhea - stool or liquid
Constipation

Causes:
- Diaper
- Decreased fiber in diet
- Decreased fluid Intake
Constipation

Impaction:
Presence of large mass of stool in colon/rectum - not moving

Treatment:
No soap suds enemas (bowel necrosis)
No Tap water enemas (electrolyte problem)

Enemas
1. Phosphate enemas - x3
2. Milk of molasses - 50% milk and molasses, osmotic diuretic
3. Oral lavage with polyethylene glycol 200-600 cc solution - (very difficult cases)
Then
4. MOM
Constipation - Treatment

Diet - ↑ Fiber
↑ Fluid

Hard stools
1. Lactulose - increases stool water content
2. Colace - increases stool water content, lubricates
3. Miralax = polyethylene glycol (pulls water)
   powder
   need fluid
   odorless, tasteless
   titrate

Frequency
1. MOM - irritant (Magnesium) GI Stimulant
2. Senna
3. Suppositories

No mineral oil - problems with aspiration
Dental Issues

Altered oral motor tone
Dysplasia
Bruxism
Tongue movement
Mouth breathing
Hypersensitive mouth
Dental Issues

Challenges to dental hygiene maintenance
Bite
Oral Aversive

Result:
- Caries
- Bad breath
- Protruding teeth
- Bleeding gums

Tx:
- Refer to Dentist - 2 years of age
- Brush
- Tongue Scrapper
- Fluoride Varnish
Sleep Problems

1. Sleep disorders in children with CP
   N = 173
   23% pathological sleep scores (5% general population)
   Difficulty in initiating and maintaining sleep
   Sleep - wake transition
   Sleep breathing disorders
   Significant: active epilepsy was associated with presence of sleep disorder
   More frequent with:
   a) Children with spastic quadraplegia
   b) Dyskinetic CP
   c) Visual impairment

Sleep Problems

Disturbances in circadian rhythm result in disturbances in sleep

Synchronized to 24º period - cues from light-dark cycle
Sleep Problems

Days/ nights mixed
  • sleeping at “wrong” times

TX: 1) Awareness - keep up
  2) Routine
  3) Medications
    Melatonin
    Amitriphyline
Sleep Problems

Melatonin

Secretion is generated by central pacemaker - “clock” - in the suprachiasmatic nuclei of the hypothalamus

Produced by pineal gland - “hormone of the darkness”

Induction of sleep
No adverse side effects
Optimum amount?
Peak - 60 minutes
Sleep Problems

Study

3 studies of Melatonin

Sleep latency (time to fall asleep) - ↓ sleep latency

Not really good studies

No differences with melatonin and placebo

1) total sleep time
2) night-time awakenings
3) parental opinions

Puberty

1. Pubic Hair
   3-4 years of age

   Early start is minimal

   Severe encephalopathy changes in brain

   Injury to pituitary hypophyseal axis - causing hormonal changes

   Premature adrenache
Puberty

2. Menstruation
   Concern
   Hygiene - Depo-Provera

3. Pregnancy
   Tease out what the concern is

4. Sex
   It happens
Future/Transition

Hard - for everyone

1. Doctor - who???

2. Education - school until 22 years of age

3. Work Programs
Child Maltreatment of Children with CP

1. Cause of Cerebral Palsy

2. Neglect
   - Overwhelmed parents
     - a. Neglected feeding with weight loss
     - b. Poor hygiene
     - c. School
     - d. Immunizations

3. Sexual Abuse

4. Usually not physical abuse
Role of Pediatrician - Team

1. Doctor
2. Medical Home = 7C’s (continuous, whole child, comprehensive, coordinated, accessible, culturally competent, family centered)
3. Team Member
4. Sounding Board/Reality
5. Cheerleader