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

Paula Kienberger Jaudes MD
University of Chicago
Pritzker School of Medicine
President and CEO
La Rabida Children's Hospital

Cerebral palsy describes a group of disorders of the development of <u>movement</u> 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.

Stevenson RD et al:Pediatrics 2006;118-3:1010-1018.

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

1. Development/Education
2. Nutrition/Growth
3. Dysphasia
4. Respiratory Issues
   - Pneumonia
   - Aspiration
   - Stridor/OSA
5. Skin Breakdown
6. Dental
7. Seizures
   - Intractable
8. Family
9. Transition
10. Neglect

Concerns are Interconnected

Parent
- Eating
- Rumination/Vomiting
- Constipation
- Too Heavy

Pediatrician
- Nutrition
- Growth
- Dysphasia
- Respiratory

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

Stallings VA, et al. *Dev Med Child Neurol* 1993;35:997-1006.
Stallings VA, et al. *Dev Med Child Neurol* 1993;35:126-138.

II. **North American Growth in CP Project**

N = 230

Moderate to Severe CP

58% - feeding problems

Fung EB, et al. *J Am Diet Assoc* 2002;102:361-368.

III. **Oxford Feeding Study**

N = 440

frequent choking = 56%

stressful and prolonged feedings = 43%

vomiting = 22%

Sullivan PB, et al. *Dev Med Child Neurol* 2002;44:461-467.

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

Rogers B. *The Journal of Pediatrics, Supplement* 2004;145(2), S28-32.

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 H_2 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

Srivastava et al. *Pediatrics* 2009;123:338-345.

Oral Aversion

Behavioral therapy works

- “Time to eat”
  - Speech Pathologist
  - Nutritionalist
  - Social Work

Can d/c GT

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)

American Academy of Pediatrics, Pediatric Nutrition Handbook, 6th Ed., 2009:831.

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.

Stevenson et al: Pediatrics 2006;118-3:1011-1018.

Indications for Gastrostomy Tube

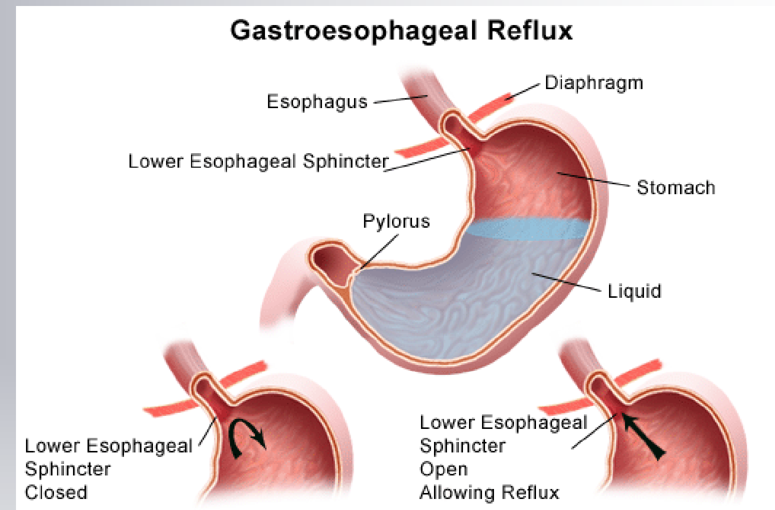
1. Dysphasia resulting in under nutrition
   - Parent's concern - heavy
2. Aspiration with associated respiratory disease
3. Insufficient fluid intake and/or refusal of oral medications
4. Excessive effort of stress during oral feeding
5. Mother - issue

Gastrostomy

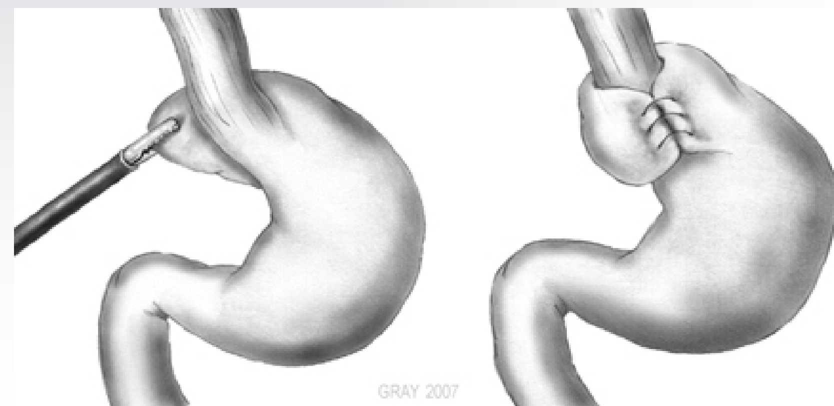
Perioperative mortality rates low

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

Little SA, et al. *Clinical Otolaryngology* 2009;34, 236-239.

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

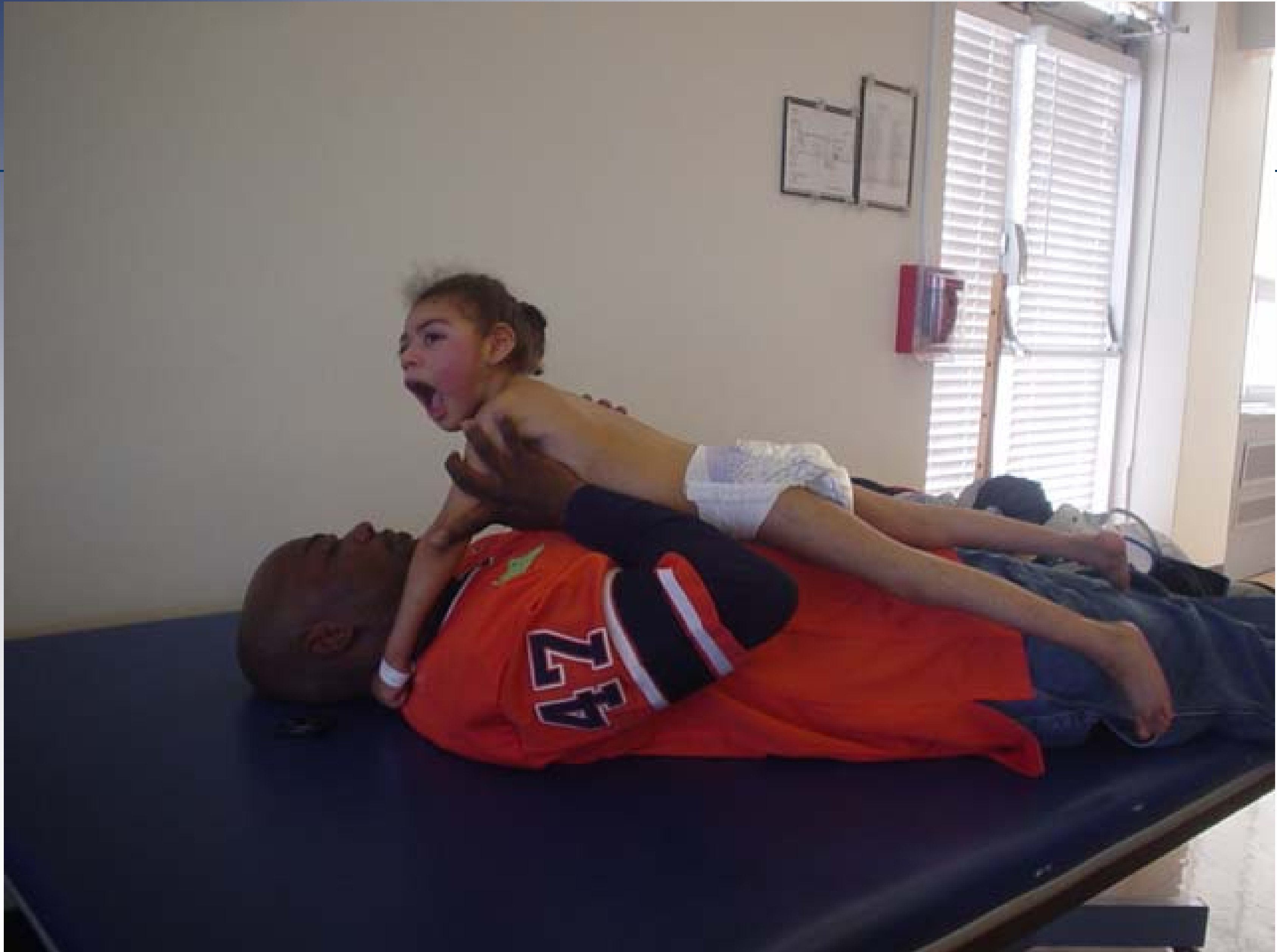
  ? Work

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: videofluroscopy – 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

Cass H et al: Developmental Medicine and Child Neurology 2005, 47:347-352.

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

Plioplys, A et al: JAMDA-September/October 2002:318-321.

Guess

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

<u>Hard stools</u>

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

<u>Frequency</u>

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

No mineral oil – problems with aspiration

- 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

Newman C et al: Developmental Medicine and Child Neurology 2006,48,564-568.

Disturbances in circadian rhythm result in disturbances in sleep

Synchronized to 24° 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

Phillips L et al: Developmental Medicine and Child Neurology 2004,46,771-775.

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
   - Educational right and responsibilities: Understanding Special Education in Illinois http://www.isbe.state.il.us/spec-ed/html/parent_rights.htm
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