

# Cerebral Palsy

## The ABC's of CP

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# Cerebral Palsy

## Outline

- I. Definition
- II. Incidence, Epidemiology and Distribution
- III. Etiology
- IV. Types
- V. Medical Management
- VI. Psychosocial Issues
- VII. Aging

# Cerebral Palsy-Definition

- Cerebral palsy is a symptom complex, (not a disease) that has multiple etiologies.
- CP is a disorder of tone, posture or movement due to a lesion in the developing brain.
- Lesion results in paralysis, weakness, incoordination or abnormal movement
- Not contagious, no cure.
- It is static, but its symptoms may change with maturation

# Cerebral Palsy

Brain damage

Occurs during developmental period

Motor dysfunction

Not Curable

Non-progressive (static)

*Any regression or deterioration of motor or intellectual skills should prompt a search for a degenerative disease*

Therapy can help improve function

# Cerebral Palsy

- There are 2 major types of CP, depending on location of lesions:
  - Pyramidal (Spastic)
  - Extrapyramidal
- There is overlap of both symptoms and anatomic lesions.

- The pyramidal system carries the signal for muscle contraction.
- The extrapyramidal system provides regulatory influences on that contraction.

# Cerebral Palsy

- Types of brain damage
  - Bleeding
  - Brain malformation
  - Trauma to brain
  - Lack of oxygen
  - Infection
  - Toxins
  - Unknown

# Epidemiology

- The overall prevalence of cerebral palsy ranges from 1.5 to 2.5 per 1000 live births.
- The overall prevalence of CP has remained stable since the 1960's.
- Speculations that the increased survival of the VLBW preemies would cause a rise in the prevalence of CP have proven wrong.
- Likewise the expected decrease in CP as a result of C-section and fetal monitoring has not happened.
- However, the prevalence of the subtypes has changed.



# Epidemiology

- Due to the increased survival of very low birth weight preemies, the incidence of spastic diplegia has increased.
- Choreoathetoid CP, due to kernicterus, has decreased.
- Multiple gestation carries an increased risk of CP.

# Distribution of the Types of CP

<b>Types of Cerebral Palsy</b>	<b>Frequency of Distribution</b>
Nonspastic (extrapyramidal and mixed types)	23%
Spastic CP (total)	77%
Spastic Diplegia	21%
Spastic Hemiplegia	21%
Spastic Quadriplegia	23%

# Etiology

- CP has multiple etiologies- many are still unknown
- Since CP is not a single entity, recurrence risks depend on the underlying cause.
- If there is a regression in skills, suspect a degenerative disease.

# Etiology

- Most causes are prenatal- genetic, congenital malformations, metabolic, intrauterine infections, rather than perinatal or postnatal- birth asphyxia, hemorrhage, infarction, infections, trauma.

# Etiology

- Much of the literature of the 1990's was directed at the controversy re the role of asphyxia in the etiology of CP
  - Asphyxia implies poor gas exchange, low Apgars and neurologic depression during and soon after delivery.
  - Significant asphyxia is accompanied by acidosis.
  - Asphyxia is rarely the cause of CP in the term infant.

# Etiology

- In one outcome study of 43,437 **full term** children, 150 had cerebral palsy. Only 9 of these cases were attributable to birth asphyxia.
- 34 had spastic quadriplegia and 71% of those cases had identifiable causes.
  - 53%- congenital disorders
  - 14%-birth asphyxia
  - 8%-CNS infections

# Etiology

- Among the children with non quadriplegic cerebral palsy, congenital disorders appeared to account for about 1/3 of the cases, and CNS infections accounted for 5%.

(Wilson and Cooley-2000; Collaborative Perinatal Study of The National Institute of Neurological and Communicative Disorders and Stroke, Naeye, 1989)

# Hypoxic Ischemic Encephalopathy (HIE)

- A clinical entity first described in 1976
- Used interchangeably with Neonatal encephalopathy.
- Asphyxia refers to the first minutes after birth (low Apgars and acidosis)
- HIE signs and symptoms persist over hours and days that follow.



# Hypoxic Ischemic Encephalopathy (HIE)

3 major lesions arise from HIE

1. Periventricular Leukomalacia (PVL) *Typically seen in the premature infant*
  - a. Hemorrhagic PVL
  - b. Ischemic PVL
2. Parasagittal Cerebral Injury  
*Typically seen in the term infant*
3. Selective (Focal) Neuronal Necrosis  
*Seen in both term and premature infants*

# Periventricular Leukomalacia (PVL)

## 1. Hemorrhagic PVL

- Hemorrhage is associated with a collection of primitive cells between the ependyma and caudate that are programmed to “melt away” at 32-34 weeks gestation
- They contain fragile capillaries that are easily damaged by hypoxia (lack of oxygen) and hypotension (drop in blood pressure).
- When the blood pressure returns to normal, bleeding occurs because the preemie has underdeveloped autoregulation.

# Periventricular Leukomalacia (PVL)

## 1. Hemorrhagic PVL (cont.)

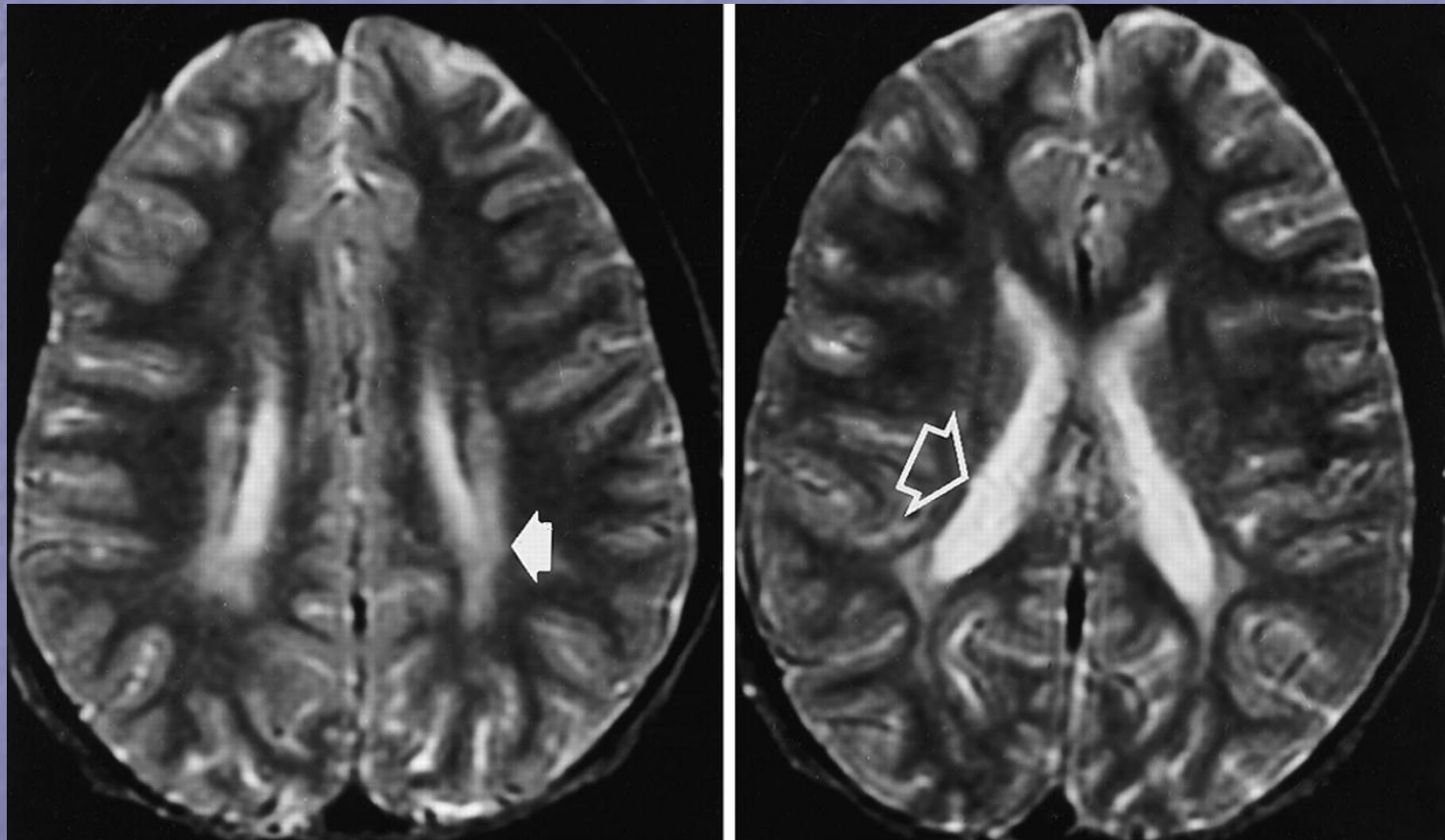
- This bleeding may then rupture into the ventricle and/or parenchyma
- Periventricular venous congestion (swelling) may then occur, and cause ischemia (lack of blood supply) and periventricular hemorrhagic infarction.

# Periventricular Leukomalacia (PVL)

## 2. Ischemic PVL

- An ischemic infarction or failure of perfusion usually to the watershed area surrounding the ventricular horns- "HIE white matter necrosis".
- Peak incidence occurs around 32 weeks
- Larger infarcts may leave a cyst
- Secondary hemorrhage can occur into these cysts- "periventricular hemorrhage".

# Periventricular leukomalacia

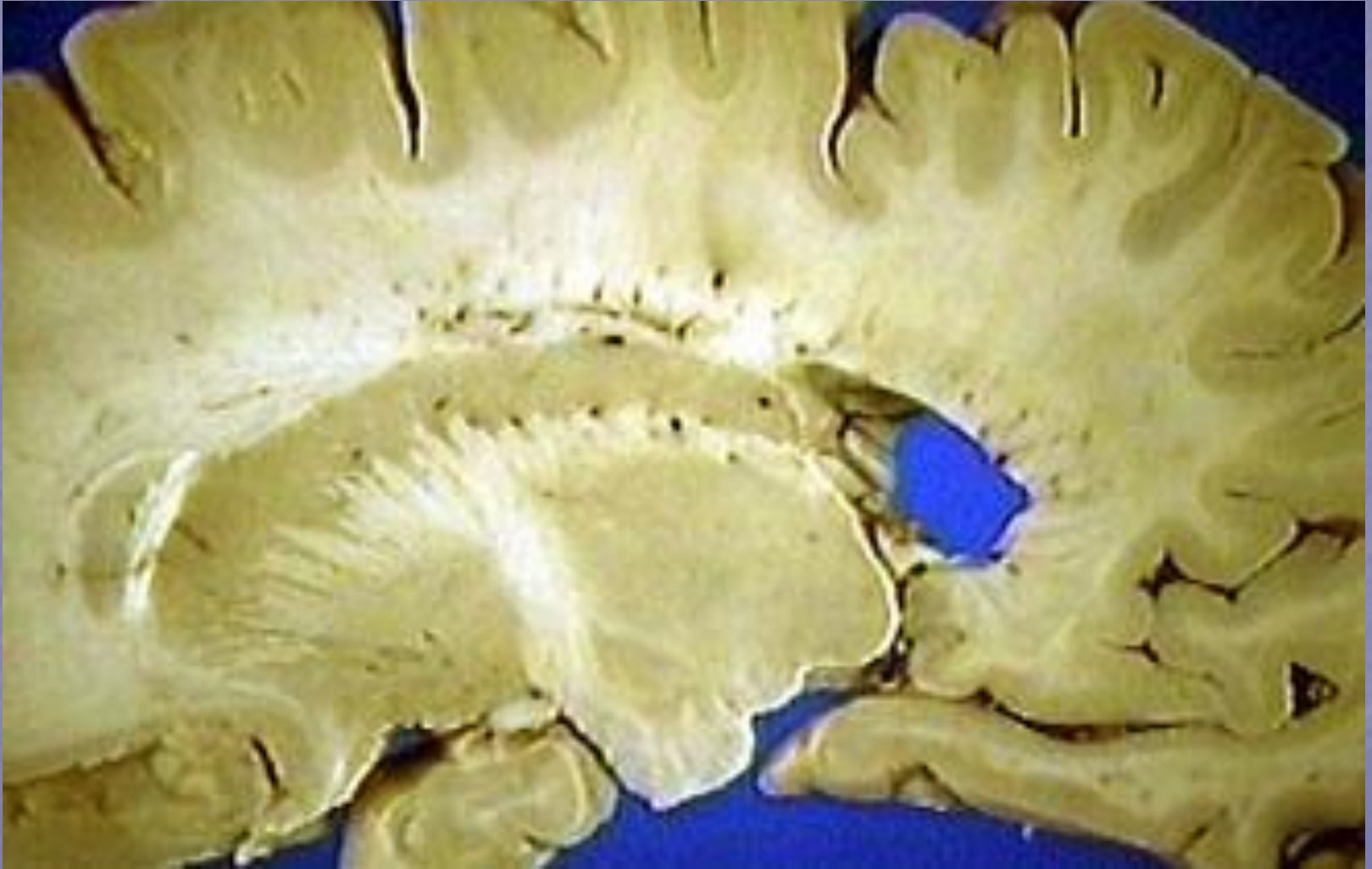


# Periventricular Leukomalacia (PVL)

## 2. Ischemic PVL

- PVL can extend into the internal capsule and result in hemiplegia superimposed on diplegia.
- Prenatal maternal ultrasound has detected lesions in the fetus at 28-32 weeks gestation, thus confirming that PVL can occur prenatally.

# Internal Capsule



# Parasagittal Cerebral Injury

- Injury is related to vascular factors, especially in the parasagittal border zones that are more vulnerable to a drop in perfusion pressure and immature autoregulation.
- The ischemic lesion results in cortical and subcortical white matter injury.
- It is usually bilateral and symmetric.
- The posterior aspect of the cerebral hemisphere especially the parietal occipital regions is more affected than the anterior.



# Selective (Focal) Neuronal Necrosis (SNN)

- Occurs in the glutamate sensitive areas in the basal ganglia, thalamus, brainstem and cortex.
- The location of the focal necrosis, which show up as cystic lesions on MRI, depend on the stage of development of the infant's brain at the time of the HIE.
  - For example, HIE at term often produces SNN in the basal ganglia since it is glutamate sensitive and very hypermetabolic at term.

# Types of Cerebral Palsy

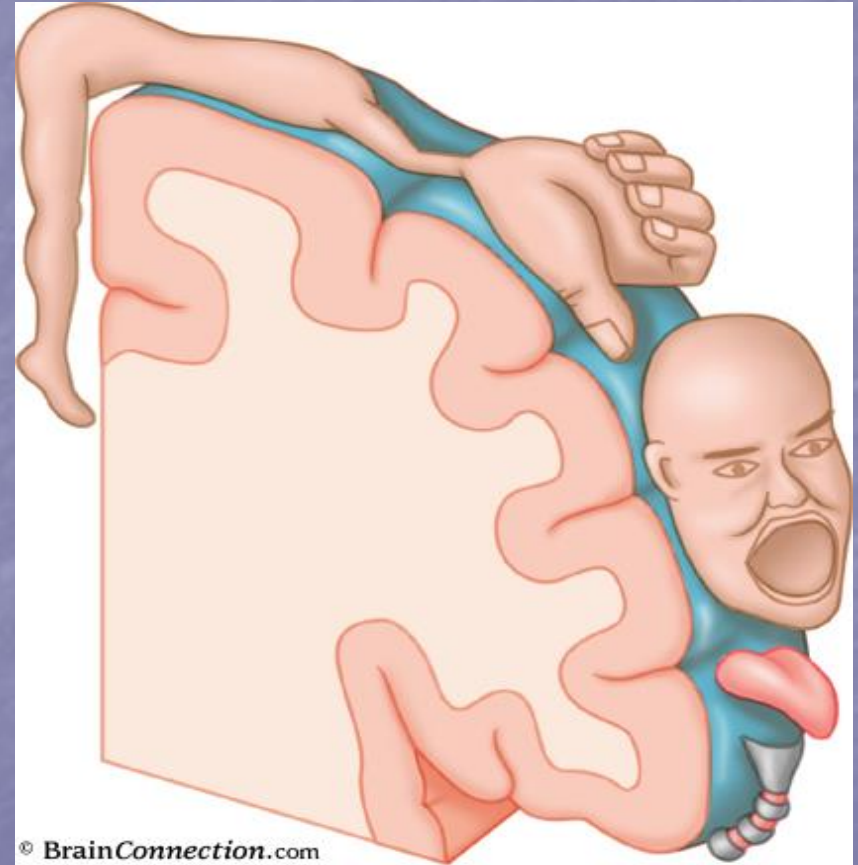
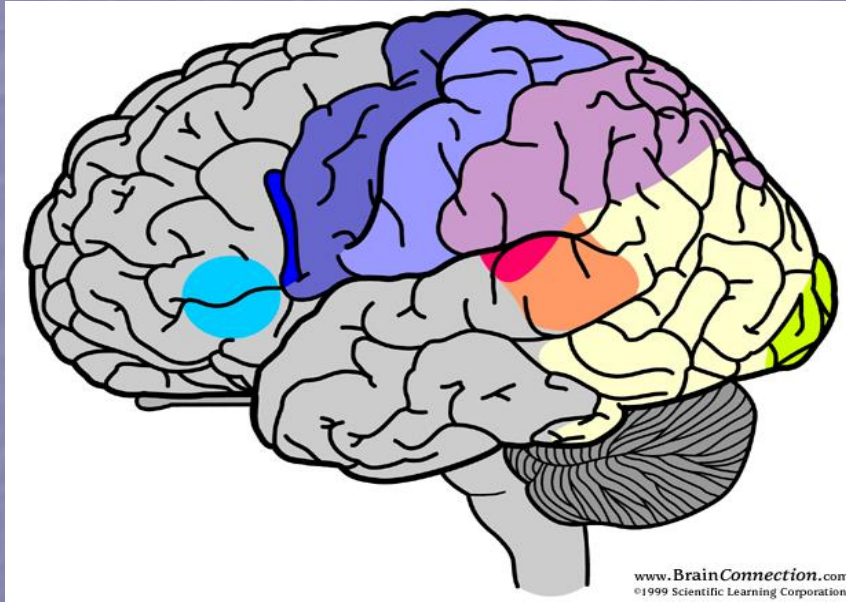
## Pyramidal

- Described as a Clasped knife response or
- Velocity dependent increased resistance to passive muscle stretch
- The spasticity can be worse when the person is anxious or ill.
- The spasticity does not go away when the person is asleep.

## Extrapyramidal

- Ataxia
- Hypotonia
- Dystonia
- Rigidity
  - The tone may increase with volitional movement, or when the person is anxious
  - During sleep the person is actually hypotonic

# Anatomy of motor lesions- pyramidal system



# Types of Cerebral Palsy

## A. **Pyramidal (Spastic)**

- Quadriplegia- all 4 extremities
- Hemiplegia- one side of the body
- Diplegia- legs worse than arms
- Paraplegia- legs only
- Monoplegia- one extremity

## **B. Extrapiramidal**

*Divided into Dyskinetic and Ataxic types*

### **Dyskinetic**

- Athetosis- slow writhing, wormlike
- Chorea- quick, jerky movements
- Choreoathetosis- mixed
- Hypotonia- floppy, low muscle tone, little movement

### **Ataxic CP**

- Results from damage to the cerebellum
- Ataxia- tremor & drunken- like gait

# Anatomy

## **Pyramidal**

- Lesion is usually in the motor cortex, internal capsule and/or cortical spinal tracts.

## **Extrapyramidal**

- Lesion is usually in the basal ganglia, Thalamus, Subthalamic nucleus and/or cerebellum.

# Comparison of Symptoms

	<b>Pyramidal</b>	<b>Extrapyramidal</b>
<b>Tone</b>	increased	alternating
<b>Type of tone</b>	spastic	rigid
<b>DTR's</b>	increased	normal to increased
<b>Clonus</b>	Present	occ. present
<b>Contractures</b>	early	late
<b>Primitive Reflexes</b>	delayed	persistent
<b>Involuntary movements</b>	rare	frequent

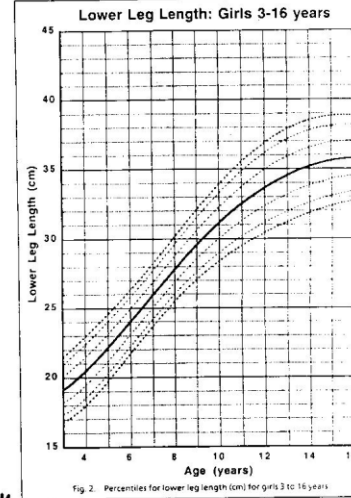
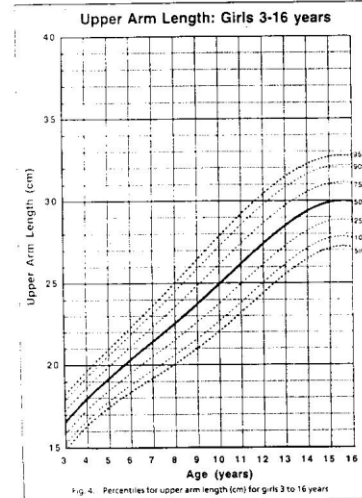
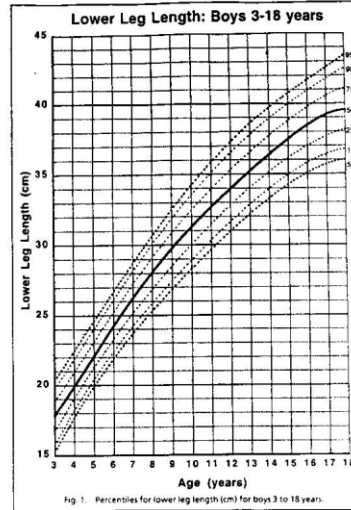
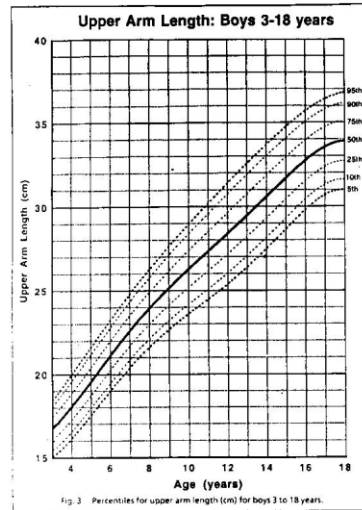
# Medical Management

## Growth

- Persons with CP often have struggle to gain or maintain weight.
- Failure to Thrive is a common problem.
  - Before diagnosing Failure to thrive, an accurate Body Mass Index must be obtained, but an accurate height is difficult to obtain in a person with severe contractures.
  - In such cases, arm span calculations may be used and a growth chart is available to determine percentiles standardized to age and gender.



# Extremity length growth chart



# Medical Management

## Orthopedic Problems

- Scoliosis
- Hip Dislocations
- Contractures
- Osteoporosis

# Medical Management

## Oromotor Dysfunction

- Especially common in persons with Extrapyrarnidal CP and Spastic quadriplegia
  - Language delay/Speech delays
  - Drooling
  - Dysphagia
  - Aspiration

# Medical Management

## Gastrointestinal Dysmotility

- Delayed gastric emptying
- Gastroesophageal reflux
  - Pain
  - Chronic aspiration
- Constipation

*These disorders are interrelated and compound one another.*

# Medical Management

## Spasticity Management

Management of spasticity does not fix the underlying pathology of CP, but it may decrease the sequelae of increased tone.

- Over time, the spasticity leads to:
  - musculoskeletal deformity
    - scoliosis
    - hip dislocation
    - contractures
  - Pain
  - Hygiene problems

# Treatment of Spasticity

## Medications

- Valium
- Dantrium
- Baclofen
- Clonidine
- Clonazepam
- BOTOX

# Treatment of Dystonia

Medications-(None are very effective)

- L-Dopa- drug of choice for certain disorders
- Artane
- Anticonvulsants-for intermittent and paroxysmal dystonia
- Anti-spasticity medications-
- Haldol or Reserpine- for choreoathetosis
- Propranolol- for essential tremor
- Clonazepam or Valium- for “rubral tremors”-(course tremors of the entire arm)
- Valproic acid or clonazepam for action myoclonus- (large jerks with intentional movements)

# Associated Problems

- Mental Retardation
- Communication Disorders
- Neurobehavioral
- Seizures
- Vision Disorders
- Hearing loss
- Somatosensation (skin sensation, body awareness)
- Temperature instability
- Nutrition
- Drooling
- Dentition problems
- Neurogenic bladder
- Neurogenic bowel
- Gastroesophageal reflux
- Dysphagia
- Autonomic dysfunction



# Other Treatments

- Casting
- Therapeutic Electrical Stimulation
- Patterning: Doman-Delacato- (*not recommended*)
- Selective Dorsal Rhizotomy
- Massage
- Hyperbaric Oxygen
- Acupuncture

# Adult Concerns

## Medical

- Routine Healthcare Maintenance
- Sequelae of Spasticity
- Orthopedic Issues
- Pain Management
- Neurogenic Bowel and Bladder
- Prevention of Chronic Aspiration Management of Gastroesophageal Reflux & Complications
  - Barrett's esophagus
  - Esophageal strictures
  - Esophageal/stomach cancer

# Adult Concerns

## Psychosocial

Transition from Pediatric to Adult services

Independence

*Work*

*Home*

Relationships

Guardianship

End of life