Developmental delay and Cerebral palsy

objectives

1. developmental delay
   - Define developmental delay
   - Etiologies of developmental delay
   - Present the differential diagnosis of developmental delay.

2. cerebral palsy
   - Definition
   - Epidemiology
   - Classification
   - Diagnosis
   - Complications
   - Prognosis
   - Management

Developmental disabilities:
   - Are a group of related chronic disorders of early onset.
   - Estimated to affect 5% to 10% of children.

**Developmental Delay**

- Global developmental delay is a subset of developmental disabilities defined as significant delay in two or more of the following developmental domains:
  - gross/fine motor
  - speech/language, cognition
  - social/personal
  - activities of daily living
- Specific developmental delay
  - Delay in a single domain
- Significant delay—defined as performance 2 standard deviations or more below the mean on age-appropriate, standardized norm referenced testing

**Developmental Surveillance**

**Examples of Screening Tests**

**Evaluation of a Child with Developmental Delay**

- **Medical Evaluation**
  - Presence of biologic risks or medical problems associated with DD
  - **Head circumference** for micro/macrocephaly
  - **Weight and height** for growth deficiency
– **Dysmorphology** (minor and major congenital abnormalities)
– **Eye exam** for poor tracking, strabismus, etc
– **Ear exam** for recurrent/chronic OM
– **Abdomen** for HSM (metabolic disease)
– **Skin** for neurocutaneous lesions
– **Neurologic exam** for reflexes, tone, symmetry, strength

• Metabolic studies (urine amino acids, serum organic acids, serum ammonia and lactate levels)
• Routine chromosome analysis
  – High resolution chromosome analysis
  – Molecular screening for subtelomeric
  – Chromosomal rDNA for fragile X testing
  – MECP 2 (Rett syndrome)
  – rearrangements (FISH)
• EEG (if history or findings on physical exam suggest epilepsy)
• MRI off brain
• Audiology evaluation
• Vision assessment

### Causes of global developmental delay

<table>
<thead>
<tr>
<th>Genetic or Syndromic</th>
<th>Identified in ~ 20% of those without neurological signs, dysmorphic features or a family history</th>
<th>Identified in ~1% of those without neurological signs, dysmorphic features or a family history</th>
</tr>
</thead>
<tbody>
<tr>
<td>Metabolic</td>
<td></td>
<td>con genital hypothyroidism</td>
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<tr>
<td>Endocrine</td>
<td></td>
<td>Acquired brain injury</td>
</tr>
<tr>
<td>Traumatic</td>
<td></td>
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<tr>
<td>Environmental Causes</td>
<td></td>
<td>This can be a contributory factor co-existing with other pathology and where the child’s needs are outside the parents’ capacity to provide for them.</td>
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<tr>
<td>Cerebral Malformations</td>
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<tr>
<td>Infections</td>
<td>• Perinatal e.g. Rubella, CMV, HIV</td>
<td>• Neonatal meningitis</td>
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<tr>
<td>Toxins</td>
<td>• Fetal: Maternal alcohol or drugs in pregnancy</td>
<td>• Kernicterus</td>
</tr>
<tr>
<td></td>
<td>• Childhood: Lead toxicity</td>
<td>• Childdoll: Lead toxicity</td>
</tr>
</tbody>
</table>
Cerebral palsy

- is a diagnostic term used to describe a group of permanent disorders of movement and posture causing activity limitation, that are attributed to non progressive disturbances in the developing fetal or infant brain.
- The motor disorders are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior as well as by epilepsy and secondary musculoskeletal problems.
- 2 to 2.5 per 1000 live births
- CDC: incidence 3.6/1000
- is the most common cause of childhood physical disability

Classification

- Topographical
- Motor abnormalities classification
  - Tone abnormality
    - Hypertonia
    - Spasticity
    - Rigidity
    - Hypotonia
  - Type of movement disorder
    - Ataxia
    - Dystonia
    - Choreaathetosis

- Etiologic
  - Prenatal (70%) Infection, anoxia, toxic, vascular, Rh disease, genetic, congenital malformation of brain
  - Natal (5-10%) Anoxia, traumatic delivery, metabolic
  - Post Natal Trauma, infection, toxic

- Functional
  Gross Motor Function Classification System (GMFCS)

Diagnosis

- Thorough
  - History
  - Physical examination
  - Preclude a progressive disorder of the NS
- degenerative diseases
- metabolic disorders
- spinal cord lesions
- muscular diseases

- The major signs that collectively can lead to a CP diagnosis are
  - delayed motor milestones
  - Abnormal neurologic examination
  - persistence of primitive reflexes
  - abnormal postural reactions.

- It is important to remember that although no single abnormal physical sign is diagnostic, clusters of symptoms or evolving abnormal movement patterns may be indications of CP and thus should be explored further

### Risk Factors Associated With Cerebral Palsy

<table>
<thead>
<tr>
<th>Prenatal</th>
<th>Perinatal</th>
<th>Postnatal</th>
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</thead>
<tbody>
<tr>
<td>Hypoxia</td>
<td>Asphyxia</td>
<td>Asphyxia</td>
</tr>
<tr>
<td>Genetic disorders</td>
<td>Premature birth &lt;32 weeks or &lt;2500 g</td>
<td>Seizures within 48 hours of birth</td>
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<tr>
<td>Metabolic disorders</td>
<td>Blood incompatibility</td>
<td>Cerebral infarction</td>
</tr>
<tr>
<td>Multiple gestation</td>
<td>Infection</td>
<td>Hyperbilirubinemia</td>
</tr>
<tr>
<td>Intrauterine infections</td>
<td>Abnormal fetal presentation</td>
<td>Sepsis</td>
</tr>
<tr>
<td>Thrombophilic disorders</td>
<td>Placental abruption</td>
<td>Respiratory distress</td>
</tr>
<tr>
<td>Teratogenic exposure</td>
<td>Instrument delivery</td>
<td>Syndrome/chronic lung disease</td>
</tr>
<tr>
<td>Chorioamnionitis</td>
<td></td>
<td>Meningitis</td>
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<tr>
<td>Maternal fever</td>
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<td>Postnatal steroids</td>
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<tr>
<td>Exposure to toxins</td>
<td></td>
<td>Intraventricular hemorrhage</td>
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<tr>
<td>Malformation of brain structures</td>
<td></td>
<td>Periventricular leukomalacia</td>
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<tr>
<td>Intrauterine growth restriction</td>
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<td>Shaken baby syndrome</td>
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<tr>
<td>Abdominal trauma</td>
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<td>Head injury</td>
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<td>Vascular insults</td>
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Data from Gibson et al., 2003; Han, Bang, Lim, Yoon, & Kim, 2002; Kuban & Leviton, 1994; Naeye et al., 1989; Nelson, 1983; Nelson & Ellenberg, 1986.

### Mental Retardation

- Not all children with CP are cognitively impaired
- there is a relationship between the severity of CP and mental retardation
- spastic quadriplegic CP have greater degrees of mental retardation than children with spastic hemiplegia.
2. **Epilepsy**
   - Up to 35% of children with CP have epilepsy
   - Onset in the first year of life in 70%
   - Focal seizures with or without secondary generalization are most common with frequently focal EEG abnormalities.
   - Children with spastic diplegic CP are at a lower risk

3. **Visual Abnormalities**
   - Children with CP, particularly preterm infants, are also at increased risk for visual impairment
     - Retinopathy of prematurity
     - Myopia
     - Strabismus
     - Glaucoma
     - Amblyopia

4. **Feeding, Nutrition, and Growth**
   - These are the most common issues encountered in children with severe CP
   - About 30% are undernourished, and many show reduced linear growth below the third percentile
   - Poor nutrition secondary to pseudobulbar palsy
   - Gastroesophageal (GE) reflux

5. **Bladder Dysfunction**
   - Increased risk for urinary incontinence, urgency, and infections
   - Spastic CP can be associated with spasticity of the detrusor muscles resulting in small frequent voids and a low capacity irritable bladder
   - Primary incontinence has been reported in up to 23% of these children and correlates with lower cognition and severe motor deficits

6. **Bowel Dysfunction**
   - Constipation is common in children with CP
   - Multiple factors including poor feeding
   - Reduced water intake
   - Immobility

7. **Sleep Disturbances**
   - Common in children with CP, particularly those with visual impairment
   - Occurring in up to 50% of cases
   - Disturbed sleep patterns with fragmented sleep
   - Frequent nocturnal awakenings

8. **Drooling**
   - Occurs in up to 30% of children with CP
• Usually secondary to mouth opening and/or swallowing difficulties due to pseudobulbar palsy

9. **Hearing Loss**
   - Certain etiologies, such as kernicterus, post-meningitis, and congenital rubella, increase the risk for hearing loss

10. **Orthopedic Abnormalities**
   - The developing bones grow in the direction of the forces placed upon them.
   - Spasticity can lead to progressive joint contractures, shortened muscles, and hip or foot deformities
   - Scoliosis and fractures due to osteomalacia or osteoporosis
   - more common with severe motor disability and immobility

**Prognosis**

• Unfortunately, it often is a "wait and see" situation

• Many patients with CP have normal intelligence but communication skills are impaired because of oro-motor, fine motor, and gross motor difficulties
  - has potential to affect the child's success both academically and socially
  - Survival and quality of life vary but are associated with the severity and number of functional disabilities

**Management**

• Pediatrician or family physician
  • pediatric physiatrists (rehabilitation)
  • developmental pediatricians
  • neurologists
  • orthopedists
  • Physical therapy
  • occupational therapy
  • speech therapy
  • adaptive equipment