Developmental Delay

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SPEECH DELAY

CEREBRAL PALSY (CP)

1. Timing of brain injury
   a. preterm
   b. early postnatal
   c. late postnatal

2. Treatment
   a. Prevention
5. Hypotonic CP
6. Ataxic CP

3. Spastic hemiparesis
4. Spastic diplegia
5. Hypotonic CP

PREVALENCE

Normal psychomotor development milestones

Learning disabilities

Developmental delay

Failure of child to achieve expected motor and cognitive milestones owing to cerebral palsy

Associated disorders:
1. Mental retardation
2. Nonprogressive encephalopathy
3. Progressive encephalopathy
4. Dyskinetic CP
5. Ataxic CP

Physical growth - decrease in size.

Development - growth in function and capability.

Development is divided into specific domains (substantial overlap exists):
1. Gross motor
2. Fine motor
3. Language (ability to understand language precedes ability to speak)
4. Cognition
5. Social/emotional growth

Most children with developmental delay are mentally retarded, and most mentally retarded children have associated handicap such as cerebral palsy or epilepsy.

The range within which normal children reach different milestones is wide - it is difficult to diagnose developmental delay within first year of life (unless it is severe).

MOTOR DEVELOPMENT

- Failure of child to achieve expected motor and cognitive milestones
- Associated disorders

Most children with expressive speech delay have normal intelligence (vs. children with accelerated speech development are often of above-average intelligence).

CEREBRAL PALSY (CP)

1. Nonprogressive motor disorder (abnormal control of movements or posture) due to intratruine or early postnatal nonprogressive (static) injury to developing brain (cerebrum or cerebellum), i.e., due to nonprogressive (static) encephalopathy.

2. Cerebral palsy does not apply to disorders of spinal cord, peripheral nerves, or muscles!

N.B. (nonprogressive) is misnomer - child’s nervous system has plasticity - some functions improve over time, others, if not treated, deteriorate.

Timing of brain injury (most important factor determining resulting pathology):

2. Second half of pregnancy - destructive injury to already formed brain.
3. Close to term or postnatal - morphology closer to term.

PREVALENCE - 2.5 children out of 1000 live births (40% are born prematurely).

DIAGNOSIS

1. Cerebral palsy
2. Spastic diplegia (diplegia), LITTLE disease
3. Spastic quadriplegia
4. Hypotonic CP
5. Dyskinetic CP

CLINICAL FEATURES

1. Manifest before age 5 yr.; symptoms may be inapparent at birth.
2. Before specific syndrome develops, symptoms include: lagging motor development and often persistent infantile reflexes, hyperreflexia, altered muscle tone.

N.B. Patients do not lose skills once acquired! (vs. progressive neurologic disorder).

3. Clinical variants (depend on lesion location):
   - Mixed CP - combination of dyskinetic CP and spastic or ataxic CP - most frequent form!! (germinal insults only rarely are specific enough to affect only one motor component).
   - Spastic hemiparesis (hemiplegia) - common form (≈ 75%) in term neonates.
   - Spastic quadraparesis (quadriplegia) - uncommon form (≈ 7%) in term neonates.
   - Hypotonic CP
   - Dyskinetic CP (athetosis, choreoathetosis) (≈ 7-20%).
8. Ataxic CP -- rarest form (~5%)

- Commonly (25%, esp. in spastic variants) associated with spectrum of developmental disabilities (mental retardation, epilepsy, visual, hearing, speech, cognitive, and behavioral abnormalities) - motor handicap may be least of child's problems.
- Spastic CP → joint contractions → joints may become misaligned.

**DIAGNOSIS**

- MRI is indicated!
- Diagnosis of underlying cause rarely influences therapy (advanced neuroimaging in CP has not been widely used until recently), but exact diagnosis is very important for parents!
- Seek for rare treatable causes (e.g. hydrocephalus)
- Vision & hearing must be tested early.

**TREATMENT**

- Parents should be taught how to handle child in daily activities (feeding, carrying, dressing, bathing, playing).
- Severe limitations in sucking and swallowing → feeding by gastroscope tube.
- Physical therapy is essential to train ambulation, stretch spastic muscles, and prevent deformities.
- Occupational therapy - self-help skills and interpersonal communication.
- Education tailored to intellectual abilities.
- Drugs / procedures for spasticity. See p. Mov1 >>

N.B. best age for spasticity surgery is 4-7 years - enough time for spontaneous improvement to occur; orthopedic problems should be fixed after spasticity is addressed!!!
- Most survive to adulthood.

**PREVENTION**

- Magnesium sulfate IV before birth for women delivering extremely premature babies.

**SPASTIC HEMIPARESIS (HEMIPLEGIA)**

- Lesion of corticospinal system of one cerebral hemisphere.

- Common causes:
  1) Intraventricular stroke (e.g. in twins, due to ischemia related to shared placental vessels); stroke can also occur during birth process and in infancy (ACUTE INFANTILE HEMIPLEGIA).
  2) Intraventricular hemorrhage (in small premature infants) complicated by intraparenchymal hemorrhage.

- Hemiparesis affects arm & hand more than leg.
- All children walk, albeit often later and on toes of affected foot (because of tight heel cord that may necessitate surgical lengthening).
- Growth "arrest" of arm and leg is frequent (esp. with parietal lobe lesions) - arm and leg are shorter and thinner, compensatory scoliosis.
- Hemiparesis may not be evident until child starts to grab for objects and shows precocious clumsiness of hands.
- Motor handicap may be least of child's problems.

**etiology**

- Most patients are premature!
- Intrauterine stroke (e.g. in twins, due to shared placental vessels);
- Adductor spasm is responsible for leg "scissoring";
- Application of diaper is difficult.
- Marked spasticity may preclude ambulation without walker and long-leg braces.
- When child is suspended by axillae, scissoring posture of lower extremities is maintained.
- Spasticity tends to increase in first and second years and is more evident when child is erect.
- Child learns to speak & read competently (speech acquisition may be delayed).
- Intelligence may be spared, but subtle neuropsychologic differences between right and left lesions may be demonstrable; 25% have cognitive abnormalities (incl. mental retardation).
- 1/3 patients have seizures (when lesion affects cortex).
- Neuroimaging - atrophic cerebral hemisphere with dilated lateral ventricle.
- Lesions:
  - Large unilateral lesion, intractable seizures, and severe behavior disorders → hemispherectomy or other excisional surgery.

**SPASTIC DIPARESIS (DIPLEGIA), s. LITTLE disease**

- Most common causes:
  1) Prematurity with bilateral germinal matrix hemorrhage ± intraventricular hemorrhage and hydrocephalus.
  - Most patients are premature!
  2) Perinatal ischemia in watershed parasagittal zone between territories of ACA and PCA.

- First noted when infant begins to crawl - child uses arms in normal reciprocal fashion but drags legs behind more as rudder (commando crawl).
- Adductor spasm is responsible for leg "scissoring";
  - Application of diaper is difficult.
- Child walks on tiptoes; marked spasticity may preclude ambulation without walker and long-leg braces.

- Joint contractures → joints may become misaligned.
- Frequent ataxia and impaired growth of lower extremities (disproportionate growth with normal development of upper torso).
- Variable clumsiness of hands.
- Intelligence and speech unimpaired.
- Likelihood of seizures is minimal.
- Neuroimaging - periventricular leukomalacia.

**SPASTIC QUADRIPLEGIA**

- Most severe variant of CP

- Often associated with moderate-to-severe mental deficiency.
- Rarely able to walk, and most are totally dependent.
- Pseudobulbar manifestations (→ aspiration pneumonia).
- Seizures are frequent.
- Poor hand use precludes learning of all but most rudimentary signs (difficult to assess cognition).
- Neuroimaging - extensive brain damage of both grey and white matter.

Spastic quadriplegia – "scissoring" of legs, pronated forearms, “fisted” hands.
HYPOTONIC CP
- floppy but with hyperactive tendon reflexes (vs. LMN or primary muscle diseases).
- pathophysiology is not understood.
- usually severe mental deficiency.

DYSKINETIC CP
- basal ganglia lesions lead to abnormal involuntary movements (athetosis, choreoathetosis, dystonia).

Chief causes:
1. Kernicterus (also see p. 1959) - unconjugated bilirubin selectively damages basal ganglia, central auditory and vestibular pathways, and deep cerebellar nuclei (cortex is not affected); may be unable to speak (because of facial dyskinesia) and hearing loss* and have little or no hand use, but may be normally intelligent; UMN signs not present, seizures uncommon.
   *hearing loss is typically in high tones; children are not deaf but cannot discriminate consonants that convey most of meaning of speech!

Yellow brain staining:

2. Severe anoxia - both cortical and subcortical damage (status marmoratus of basal ganglia) → intellectual as well as motor handicaps.
   - movements emerge after age 1 year (in early infancy, children are hypotonic, with poor head and trunk control and little or no use of hands) - first sign may be tongue thrusting (makes spoon feeding difficult).
   - some children walk but assume unusual postures and have stigmatizing facial grimaces, dysarthria, and dysphagia.

Treatment - high doses of TRIXYPHENIDE, LEVODOPA, CARBAMAZEPINE may have modest effect; stereotactic surgery on basal ganglia and thalamus runs risk of irreversible anarthria (if lesions impinge on internal capsule).

ATAXIC CP
- rarest form, due to maldevelopment of cerebellum or its pathways.
   - truncal and gait ataxia are more striking than limb ataxia, but some children take long time to learn to feed themselves and have severe difficulty writing.
   - eventually learn to walk but remain clumsy and fall frequently.
   - nystagmus is uncommon.
   - speech may be slow and scanning.
   - neuroimaging - most have no abnormal findings (only 25% have detectable posterior fossa pathology; small number - supratentorial malformations).
   - may improve with age.

BIBLIOGRAPHY for ch. “Pediatrics” → follow this LINK >>