

Developmental Delay

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NORMAL PSYCHOMOTOR DEVELOPMENT MILESTONES → see p. D5 >>	
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PHYSICAL GROWTH - increase 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
- progress within different domains varies.

Developmental delay - **failure of child to achieve expected motor and cognitive milestones** owing to **ENCEPHALOPATHY**.

Associated disorders:

- 1) **mental retardation**
- 2) **nonprogressive encephalopathy** - previous brain injury that is no longer active → *STATIC DISORDERS OF BRAIN DEVELOPMENT* (e.g. cerebral palsy)
- 3) **progressive encephalopathy** (expanding mass lesion, neurometabolic, neurodegenerative or chronic inflammatory diseases)
- 4) **spinal dysraphism**
- 5) **autism**.

Most children with developmental delay are mentally retarded, and most mentally retarded children have associated handicap such as cerebral palsy or epilepsy.
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- 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* cannot be significantly accelerated by applying increased stimulation.
- appropriate attachments and nurturing in infancy and early childhood are critical factors in *COGNITIVE* and *EMOTIONAL GROWTH*.
- it may be very difficult to discriminate between *motor delay* and *mental retardation*.

SPEECH DELAY

- delays in **expressive speech** are typically not accompanied by other developmental delays (vs. delays in **both receptive and expressive speech** - often additional developmental problems).
- evaluation should start with **hearing assessment**.
Hearing deficits impair language development (hearing problems must be remedied as early as possible!!!)
- most children who experience speech delay have *normal intelligence* (vs. children with accelerated speech development are often of *above average intelligence*).

CEREBRAL PALSY (CP)

- **nonprogressive motor disorder (abnormal control of movements or posture)** due to **intrauterine ÷ early postnatal** nonprogressive (static) injury to DEVELOPING brain (**cerebrum** or **cerebellum**), i.e. due to **nonprogressive [static] encephalopathy**.

N.B. term "cerebral palsy" does not apply to disorders of spinal cord, peripheral nerves, or muscles!

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

- a) **early fetal life** → arrested / altered development of immature brain → congenital malformation.
- b) **second half of pregnancy** → destructive injury to already formed brain.
- c) **close to term or postnatal** → morphology closer to adult pathology.

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

ETIOLOGY

- cause could not be identified in most cases!
- **intrauterine** (90%) / **neonatal** (10%) factors that **injure DEVELOPING brain**:
 1. Ischemic / anoxic accidents
 2. Malformations
 3. Infections
 4. Kernicterus

CLINICAL FEATURES

- manifest before age 5 yr.; symptoms may be inapparent at birth.
- 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 disorders!)
- **clinical variants** (depend on lesion location):
 1. Spastic diparesis (diplegia), **LITTLE disease** – most common form (≈ 45%).
 2. Spastic hemiparesis (hemiplegia) – commonest form (≈ 34%) in term neonates.
 3. Spastic quadriplegia (quadriplegia) (≈ 7%)
 4. Hypotonic CP
 5. Dyskinetic CP (athetosis, choreoathetosis) (≈ 7-20%)
 6. Ataxic CP – rarest form (< 5%)
 7. Mixed CP – combination of dyskinetic CP and spastic or ataxic CP.
- 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 contractures → 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 **gastrostomy tube**.
- **physical therapy** is essential to train ambulation, stretch spastic muscles, and prevent deformities.
- **drugs / procedures** for spasticity. see p. Mov3 >>
- **occupational therapy** - self-help skills and interpersonal communication.
- **education** tailored to intellectual abilities.
- 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) **intrauterine 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 handedness or failure of hand use; this does not imply that lesion was acquired postnatally.
- 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.
- treatment:
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 prematures!
 - 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.
 - when child is suspended by axillae, scissoring posture of lower extremities is maintained.
- disuse atrophy 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.

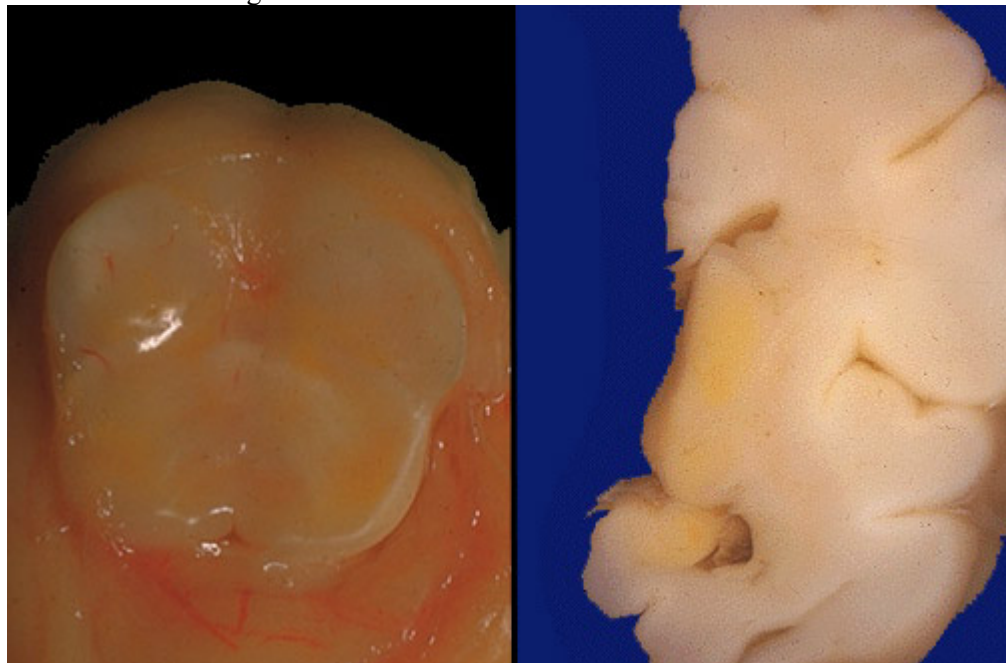
DYSKINETIC CP

- 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 **TRIHEXYPHENIDYL**, **LEVODOPA**, **CARBAMAZEPINE** may have modest effect; **stereotaxic 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 >>](#)