Developmental Delay

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**Normal Psychomotor Development Milestones** → see [p. D5 >>](http://www.neurosurgeryresident.net/D.%20Diagnostics\D1-5.%20Neurologic%20Examination\D5.%20Pediatric%20Neurologic%20Examination.pdf#Psychomotor_development)

**Learning Disabilities** → see [p. Psy31 >>](http://www.neurosurgeryresident.net/Psy.%20Psychiatry\Psy31.%20Learning%20Disabilities.pdf)

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 **nonprogressive encephalopathy** - previous brain injury that is no longer active → *static disorders of brain development* (e.g. cerebral palsy)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.

* + 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!

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):

1. **early fetal life** → arrested / altered development of immature brain → congenital malformation.
2. **second half of pregnancy** → destructive injury to already formed brain.
3. **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. **Mixed CP** – combination of dyskinetic CP and spastic or ataxic CP – most frequent form!!!! (perinatal insults only rarely are specific enough to affect ony one motor component)
2. **Spastic diparesis (diplegia),** **Little disease** – most common form (≈ 45%).
3. **Spastic hemiparesis (hemiplegia)** – commonest form (≈ 34%) in term neonates.
4. **Spastic quadriparesis (quadriplegia)** (≈ 7%)
5. **Hypotonic** CP
6. **Dyskinetic** CP (athetosis, choreoathetosis) (≈ 7-20%)
7. **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 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.
  + **occupational therapy** - self-help skills and interpersonal communication.
  + **education** tailored to intellectual abilities.
  + **drugs / procedures** for spasticity. [see p. Mov3 >>](http://www.neurosurgeryresident.net/Mov.%20Movement%20disorders,%20Ataxias\Mov3.%20GENERAL%20-%20UMN%20(pyramidal)%20&%20LMN%20Disorders.pdf#Treatment_of_spasticity)

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. 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!

1. 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

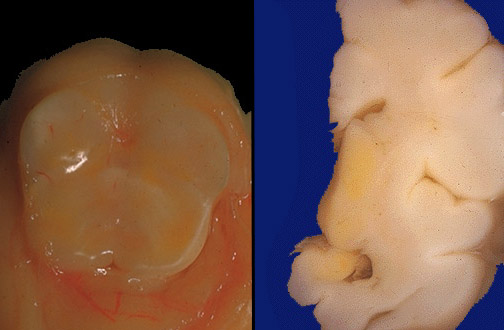
- basal ganglia lesions lead to abnormal involuntary movements (athetosis, choreoathetosis, dystonia).

Chief causes:

1. **Kernicterus** (also see [p. 1959 >>](http://www.neurosurgeryresident.net/USMLE%202\Digestive%20system%20(1801-2050)\1959%20(8a).%20Cholestasis.pdf)) - 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:



1. **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 >>](http://www.neurosurgeryresident.net/Ped.%20Pediatrics\Ped.%20Bibliography.pdf)

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