Developmental Delay

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**Normal Psychomotor Development Milestones** → see [p. D5 >>](http://www.neurosurgeryresident.net/D.%20Diagnostics%5CD1-5.%20Neurologic%20Examination%5CD5.%20Pediatric%20Neurologic%20Examination.pdf#Psychomotor_development)

**Learning Disabilities** → see [p. Psy31 >>](http://www.neurosurgeryresident.net/Psy.%20Psychiatry%5CPsy31.%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 **nonprogressive encephalopathy** - previous brain injury that is no longer active → *static disorders of brain development* (e.g. cerebral palsy)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%2C%20Ataxias%5CMov3.%20GENERAL%20-%20UMN%20%28pyramidal%29%20%26%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%5CDigestive%20system%20%281801-2050%29%5C1959%20%288a%29.%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%5CPed.%20Bibliography.pdf)

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