McCance: Pathophysiology, 6th Edition

Chapter 19: Alterations of Neurologic Function in Children

Key Points – Print

SUMMARY REVIEW

Structure and Function of the Nervous System in Children

1. The central nervous system develops from the neural tube, which is ectodermal in origin. The cranial end of the tube forms the brain, and the spinal cord is formed from the remainder of the tube.
2. The cranial and spinal ganglia (peripheral nervous system) develop from the neural crest.
3. The nervous system develops in six stages, and disruption of any of the stages can lead to malfunction of the nervous system.
4. The bones of the skull are joined by sutures; the wide, membranous junctions of the sutures, known as *fontanels*, close by 20 months of age.
5. Myelin is a sheath that develops around axons to facilitate speed of nerve impulse conduction. Progressive development of reflexes corresponds to normal maturation of nerve tissue.
6. Neurologic functioning at birth is at the subcortical level with reflex patterns mediated by the brainstem and spinal cord. With maturation, neonatal reflexes disappear and voluntary motor functions develop.
7. Head circumference is one fourth of the total height in infants compared with one eighth in adults. The fontanels allow for cranial expansion because the head is the fastest growing body part during infancy.

Structural Malformations

1. Defects of neural tube closure include anencephaly (absence of part of the brain and soft, bony part of skull) encephalocele (protrusion of brain and meninges through a skull defect), meningocele (cystlike defect with protrusion of spinal fluid–filled meninges through a vertebral defect), and myelomeningocele (a defect like meningocele only also containing the spinal cord).
2. Failure of the vertebrae to close with protrusion of neural tube contents is known as *spina bifida*.
3. Acrania is nearly complete absence of the cranial vault.
4. Premature closure of the cranial sutures causes craniosynostosis and prevents normal skull expansion and compression of growing brain tissue.
5. Microcephaly is lack of brain growth and retarded mental and motor development.
6. Congenital hydrocephalus results from an imbalance between the production and reabsorption of cerebrospinal fluid.

Encephalopathies

1. Static encephalopathies (i.e., cerebral palsy and epilepsy) are nonprogressive disorders of the brain that can occur during gestation, birth, or childhood and can be caused by endogenous or exogenous factors.
2. Cerebral palsy is a group of nonprogressive syndromes that can be caused by prenatal cerebral hypoxia or perinatal or postnatal trauma with symptoms of mental retardation, seizure disorders, or developmental disabilities.
3. Inherited metabolic disorders that damage the nervous system include defects in amino acid metabolism (phenylketonuria) and lipid metabolism (Tay-Sachs disease) and result in abnormal behavior, seizures, and deficient psychomotor development.
4. Seizures are the abnormal discharge of electrical activity within the brain. Seizure disorders are associated with numerous nervous system disorders and more often are a generalized rather than a partial type of seizure. Epilepsy is recurrence of seizure activity.
5. Generalized forms of seizures include tonic-clonic, myoclonic, atonic, akinetic, and infantile spasms.
6. Partial seizures suggest more localized brain dysfunction.
7. Febrile seizures usually are limited to the ages of 9 months to 3 years, with a pattern of one seizure per febrile illness.
8. Reye syndrome is an acute encephalopathy associated with influenza B and varicella viruses and symptoms of hypoglycemia, hyperammonemia, and increased serum short-chain fatty acids. Progressive manifestations include lethargy, stupor, rigidity, seizures, and respiratory arrest.
9. Accidental poisonings from a variety of toxins can cause serious neurologic damage.
10. Bacterial meningitis is commonly caused by *H. influenzae, N. meningitidis,*or *S. pneumoniae* and may result from respiratory or gastrointestinal infections with symptoms of fever, headache, photophobia, seizure, rigidity, and stupor.
11. Viral meningitis presents similar to bacterial meningitis, and the specific virus is often unknown.
12. HIV-1 encephalopathy is a CNS infection that can occur in infants and children.

Cerebrovascular Disease in Children

1. Occlusive cerebrovascular disease may result from embolism, sinovenous thrombosis, or congenital or iatrogenic vessel narrowing.
2. Congenital arteriovenous malformations are the most common cause of intracranial bleeding and hemorrhagic stroke in children.

Childhood Tumors

1. Brain tumors are the most common tumors of the nervous system and the second most common type of childhood cancer.
2. Tumors in children are most often located below the tentorial membrane.
3. Fast-growing tumors produce symptoms early in the disease, whereas slow-growing tumors may become very large before symptoms appear.
4. Symptoms of brain tumors may be generalized or localized. The most common general symptom is increased ICP (headache, irritability, vomiting, somnolence, and bulging of fontanels).
5. Localized signs of infratentorial tumors in the cerebellum include impaired coordination and balance. Cranial nerve signs occur with tumors near the brainstem.
6. Supratentorial tumors may be located near the cortex or deep in the brain. Symptoms depend on the specific location of the tumor.
7. Neuroblastoma is an embryonal tumor of the sympathetic nervous system and can be located anywhere there is sympathetic nervous tissue. Symptoms are related to tumor location and size of metastasis.
8. Retinoblastoma is a congenital eye tumor that has a hereditary and a nonhereditary form.