

# Pediatric Neuroimaging

# FIFTH EDITION

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To our families, without whose inspiration none of this would have been possible.

## PREFACE TO THE FIRST EDITION

ew techniques for pediatric brain and spine diagnoses have rapidly developed over the past ten years. Computed tomography, ultrasound, and magnetic resonance imaging have opened a new window to the pediatric central nervous system. Through the use of these imaging modalities, an increased understanding of the pathological processes that occur in the pediatric brain has emerged. However, in spite of the wealth of new concepts that have evolved from these new resources, there has been a notable lack of textbooks on the subject, particularly in dealing with CT and MR. In this book, I attempt, at lease in part, to fill the gap of knowledge that exists in pediatric neuroimaging.

This book strongly emphasizes CT and MR in pediatric neurodiagnosis. The reasons for this are twofold. First, there are a number of good textbooks available that focus on plain film and sonographic evaluation of the pediatric central nervous system. Second, and more important, I feel that CT and MR, particularly MR, are the best modalities by far for imaging the pediatric brain. In those areas where ultrasound and plain film radiology are important adjuncts or are of primary importance in diagnosis, they have been included. Specifically, this includes the diagnosis of intracranial pathology in premature infants.

Readers will note that this is not an encyclopedic work on diseases of the pediatric central nervous system. Those disease processes that are well covered in other texts, or are extremely uncommon, are deemphasized here. Instead, I have attempted to cover subjects that are encountered in everyday practice. Furthermore, I have emphasized concepts that are crucial to proper imaging techniques and image interpretation. Embryology, normal development, and pathophysiology are explained. Once these basic concepts are understood, interpretation of images is greatly facilitated. Finally, an attempt was made to present the information in a concise and straightforward manner that will make reading this book an enjoyable learning experience.

## PREFACE

he rapid evolution of Pediatric Neuroimaging continues to accelerate. New imaging techniques continue to emerge and improve to the point where they become useful in the imaging evaluation of children. New diseases continue to be described, along with new categories of diseases and new ways to classify them. In addition, the diseases are better understood, as a result of advances in genetics, molecular biology, biochemistry, and imaging. It has become difficult to keep up with all of it; thus, for the first time, it no longer seemed feasible for a single author to update the entire book. As a result, the fifth edition of Pediatric Neuroimaging is a collaborative effort. Charles Raybaud and James Barkovich have written much of it and edited the work of seven other authors (Christopher Hess, Pratik Mukherjee, Zoltan Patay, Erin Schwartz, Gilbert Vezina, Gary Hedlund, and Phil Meyers), who have graced this edition with information from their areas of expertise. The result is a more complete book, which shares the experience and opinions of dedicated pediatric neuroradiologists with many decades of experience in many different locations. Contributions by many authors increase the risk of an uneven text, with much overlap, differences in style, and the potential for contradictions. The authors, indeed, have different styles, but the book was edited to minimize those differences and, at the same time, keep the book readable so that it can be used as a tool for learning, as well as a reference text. In all chapters, the readers will find extensive use of tables to help to organize the disorders and images to illustrate them.

Chapter 1 on imaging techniques and Chapter 2 on normal developmental features seen by imaging have been updated with attention to new information on limiting exposure to ionizing radiation while discussing both more traditional and new imaging techniques (diffusion tractography, arterial spin labeling, and volumetrics) and protocols for MRI. Chapter 2 also has a new section on resting state functional MRI, potentially a very interesting and useful technique.

Readers will note that, after Chapter 2, all new techniques have been integrated into the discussion of specific disease entities, which is still organized by category of disease. Thus, fetal images, perfusion studies, diffusion imaging, PET images, or proton spectroscopy are illustrated when they are useful for diagnosis. For malformations, hydrocephalus, and brain injury, in particular, fetal imaging is extremely useful for early detection of disease. These fetal images show all of the same features of these disorders that are identified in infants and children and can be identified using the same search patterns; there is no reason to put them in a separate section.

Chapter 3 on metabolic diseases has been expanded to include many newly described disorders and groups of disorders. It also includes a new section on genetic disorders with extensive cerebellar involvement (particularly cerebellar hypoplasia and cerebellar atrophy) as the major imaging finding. In addition, new information on diffusivity, spectroscopy, and clinical/genetic manifestations has been added. We have retained the two section approach, with the initial section having a brief, diagnosisoriented discussion and the longer second section giving a more detailed explanation of the disorders and their imaging manifestations.

Chapter 4 on destructive lesions of the brain and spine includes new information on imaging of pediatric stroke, imaging of brain injury in preterm and encephalopathic term neonates (including birth asphyxia), as well as traumatic brain injury (accidental and inflicted, both of which continue to be topics of hot debate). We have tried to give a fair and balanced view of controversial issues.

Chapter 5 on malformations has been extensively updated with newly described disorders, as well as new information, theories, genetics, and classifications. The organization of the chapter has changed; it is now organized according to the part of the brain primarily involved by the disease process: (dorsal forebrain [cerebral cortex and commissures], ventral forebrain [base of the brain], midbrain/hindbrain, craniocervical junction, brain coverings) in order to help the reader locate the disorder within the text; we believe that this will be useful to readers. Chapters 6 (Phakomatoses) and 7 (Brain Tumors) have been updated with new genetic information, new classifications, many new images, and several new disorders being described.

Chapter 8 on hydrocephalus has been significantly modified and upgraded to include new theories of hydrocephalus that better explain the effects upon the ventricular system and the underlying brain. Chapters 9 (Spine Anomalies), 10 (Spine Tumors), and 12 (Disorders of Cerebral Blood Vessels) have all been carefully updated with new data, new theories, and new images that facilitate the diagnosis and understanding of these disorders. Chapter 11 (Infection) has been greatly expanded and improved by adding many new disorders and images, in order to expand our understanding of viruses, disorders that are prevalent outside of North America and Europe, and newly described diseases.

Despite the many changes in this edition, we hope the reader will notice that the philosophy of the book remains the same. A large number of disorders are discussed and illustrated, as it is much easier to recognize a disorder by seeing imaging than by reading about imaging features. The cause of the disorder, the main clinical features, and the underlying cause/pathophysiology are discussed whenever possible because it is easier to remember disorders when the genetic or embryologic or destructive cause is understood, rather than trying to match imaging characteristics to a disease name.

For the convenience of the reader, some topics are discussed more than once in the text. The purpose of this is to avoid forcing the reader to page back through the book, trying to find the previous mention of a disorder. For example, Chiari II malformations are discussed in Chapter 5, under disorders of the craniocervical junction, as a brain malformation and also in Chapter 9 under myelomeningoceles because they are almost always associated. Within Chapter 5, disorders secondary to abnormal pial basement membrane formation are discussed in both the dorsal forebrain section and the midbrain/ hindbrain section because both regions are variably involved in the pathologic process, such that they might present as a forebrain or a hindbrain malformation.

We hope that this new edition of *Pediatric Neuroimaging* will serve as a textbook for residents, fellows, and practicing physicians who are interested in diseases of the pediatric brain and spine while, at the same time, serving as a reference book for clinicians seeing patients with these diseases in their daily work.

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## LIST OF DISORDERS

## CHAPTER 3: METABOLIC, TOXIC, AND INFLAMMATORY BRAIN DISORDERS

## IV.B. Metabolic Disorders Primarily Affecting White Matter

- 1. White matter diseases initially affecting periventricular cerebral white matter
  - a. Metachromatic leukodystrophy
  - b. Globoid cell leukodystrophy (Krabbe disease)
  - c. Classic X-linked adrenal leukodystrophy/adrenomyeloneuropathy/acyl-CoA-oxidase deficiency
  - d. Leukoencephalopathy with vanishing white matter
  - e. Giant axonal neuropathy
  - f. Phenylketonuria
  - g. Maple syrup urine disease
  - h. Hyperhomocysteinemia (formerly known as homocystinuria)
  - i. Cystathionine beta synthase deficiency
  - j. 5, 10 methylenetetrahydrofolate reductase deficiency (MTHFRD)
  - k. Errors affecting cobalamin (vitamin B<sub>12</sub>) metabolism
  - l. Biotinidase deficiency
  - m. Methionine adenosyltransferase deficiency
  - n. Oculocerebrorenal syndrome (Lowe syndrome)
  - o. Merosin-deficient congenital muscular dystrophy (MDC1A)
  - p. Mucolipidosis type IV
  - q. Autosomal recessive spastic paraplegia with thin corpus callosum
  - r. Sjögren-Larsson syndrome
  - s. Brain injury from radiation and chemotherapy
- 2. White matter disorders with dysmyelination initially affecting subcortical cerebral white matter
  - a. Megalencephalic leukoencephalopathy with subcortical cysts (MLC)
  - b. Cystic leukoencephalopathy without megalencephaly
  - c. Aicardi-Goutières syndrome
  - d. Cockayne syndrome
  - e. Galactosemia
- 3. White matter disorders due to hypomyelination (hypomyelinating leukodystrophies)
  - a. Pelizaeus-Merzbacher disease
  - b. Pelizaeus-Merzbacher-like disease
  - b. Pelizaeus-Merzbacher-like disease
  - c. Leukodystrophies with trichothiodystrophy
  - d. 18q-Syndrome and other chromosome 18 mutations
  - e. Sialuria
  - f. Hypomyelination with congenital cataracts
  - g. Fucosidosis
  - h. Hypomyelination with atrophy of the basal ganglia and cerebellum (HABC)

- 4. White matter diseases with nonspecific patterns
  - a. Nonketotic hyperglycinemia (glycine encephalopathy)
  - b. Dihydropyrimidine dehydrogenase deficiency
  - c. 3-Hydroxy-3-methylglutaryl-coenzyme A lyase deficiency
  - d. Congenital white matter hypoplasia/familial spastic paraplegia
- 5 Idiopathic inflammatory, autoimmune, infectious, and toxic disorders affecting white matter
  - a. Multiple sclerosis
  - b. Neuromyelitis optica (Devic disease)
  - c. Acute disseminated encephalomyelitis (ADEM)
  - d. Acute hemorrhagic encephalomyelitis
  - e. Collagen vascular diseases/systemic lupus erythematosus
  - f. Osmotic myelinolysis in childhood
  - g. Toxins
  - h. Lead encephalopathy
  - i. Solvent abuse
  - j. Progressive multifocal leukoencephalitis

#### IV.C. Metabolic Disorders Primarily Involving Gray Matter

- 1. Gray matter disorders primarily involving the deep gray cerebral nuclei
  - a. Pantothenate kinase-associated neuropathy (neurodegeneration with brain iron accumulation 1, formerly Hallervorden-Spatz disease)
  - b. Juvenile Huntington disease
  - c. Isovaleric acidemia
  - d. Succinic semialdehyde dehydrogenase deficiency
  - e. Creatine deficiency syndromes
  - f. Wernicke encephalopathy
  - g. Extrapontine myelinolysis
  - h. Hemolytic-uremic syndrome
  - i. Sydenham chorea
  - j. Chronic liver disease
- 2. Gray matter disorders primarily involving cortex
  - a. Neuronal ceroid lipofuscinosis
  - b. Aspartylglucosaminuria
  - c. Anti-N-methyl-D-aspartate receptor (NMDAR) encephalitis in children and adolescents
  - d. Infantile neuroaxonal dystrophy
  - e. Niemann-Pick disease
  - f. Rett syndrome
  - g. Toxins
- h. Progressive cerebral poliodystrophy (Alpers disease)

# IV.D. Metabolic Disorders that Affect both Gray and White Matter

- 1. Canavan disease (aspartoacylase deficiency, spongiform leukodystrophy)
- 2. Fibrinoid leukodystrophy (Alexander disease)

- 3. Mucopolysaccharidoses
- a. Multiple sulfatase deficiency
- 4. Peroxisomal disorders
  - a. Peroxisomal biogenesis disorders
  - b. Acyl-CoA-oxidase deficiency
  - c. Rhizomelic chondrodysplasia punctata
  - d. Nonrhizomelic chondrodysplasia punctata
- 5. Wilson disease
- 6. Mitochondrial disorders (respiratory chain disorders)
  - a. Mitochondrial encephalomyopathy with lactic acidosis and strokelike episodes (MELAS)
  - b. Kearns-Sayre syndrome/progressive external ophthalmoplegia
  - c. Mitochondrial neurogastrointestinal encephalomyopathy
  - d. Disorders causing Leigh syndrome
  - e. Alpers disease
  - f. Trichopoliodystrophy (Menkes disease)
  - g. Glutaric aciduria type I (glutaryl-CoA-dehydrogenase deficiency)
  - h. Glutaric aciduria type II (multiple acyl-CoA-dehydrogenase deficiency)
  - i. Neonatal lactic acidosis, complex I/IV deficiency, and fetal cerebral disruption
  - j. Friedreich ataxia
  - k. Ethylmalonic encephalopathy
  - l. Nonspecific mitochondrial disorders
- 7. Pyruvate dehydrogenase deficiency
- 8. Disorders of the urea cycle and ammonia
- 9. Methylmalonic and propionic acidemias
- 10. GM1 and GM2 (Tay-Sachs and Sandhoff diseases) gangliosidoses
- 11. Fucosidosis
- 12. l-2-Hydroxyglutaric aciduria
- 13. Acute necrotizing encephalitis
- 14. Hypomyelination with atrophy of the basal ganglia and cerebellum
- 15. 3-Methylglutaconic aciduria and Barth syndrome
- 16. Molybdenum cofactor deficiency
- 17. Isolated sulfite oxidase deficiency
- 18. Toxin ingestions

#### IV.E. Metabolic Disorders Primarily Involving the Cerebellum

- 1. Friedreich ataxia
- 2. Ataxia-Telangiectasia
- 3. Late onset GM2 gangliosidosis
- 4. Ataxia with oculomotor apraxia, types 1 and 2
- 5. Autosomal recessive spastic ataxia (of Charlevoix-Saguenay)
- 6. Mitochondrial disorders causing cerebellar atrophy a. Coenzyme Q<sub>10</sub> deficiency
  - b. SANDO syndrome
  - c. Infantile onset spinocerebellar ataxia
- 7. Infantile olivopontocerebellar atrophy
- 8. Pontocerebellar hypoplasia
- 9. Congenital disorders of glycosylation
- 10. Mevalonic kinase deficiency (Mevalonic aciduria)
- 11. Progressive encephalopathy with edema, hypsarrhythmia, and optic atrophy (PEHO)
- 12. Dentatorubral and pallidoluysian atrophy
- 13. Neuronal ceroid lipofuscinoses
- 14. Langerhans cell histiocytosis
- 15. Hypomyelination with atrophy of the basal ganglia and cerebellum (HABC)
- 16. Spinocerebellar ataxias

- 17. Cerebrotendinous xanthomatosis
- 18. Wolfram syndrome
- 19. Marinesco-Sjögren syndrome
- 20. X-linked nonprogressive congenital cerebellar hypoplasia
- 21. Höyeraal-Hreidarsson syndrome
- 22. Revesz syndrome

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- 2. Hydranencephaly
- 3. Encephalomalacia

#### III. Hypoxic-ischemic Brain Injury

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  - a. Manifestations and causes of infarctions in children
  - b. Choice of radiologic study in pediatric stroke
  - c. Arterial infarctions Presumed perinatal ischemic stroke Transient cerebral arteriopathy
  - d. Infarction secondary to venous occlusion
- 2. Diffuse ischemic or inflammatory brain injury
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    - Profound hypotension in premature neonates
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    - Profound hypotension
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- VII. Brain Injury Associated with Congenital Heart Disease

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#### IX. CNS Trauma in Infancy and Childhood

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  - a. Spinal cord injury
  - b. Nerve root and brachial plexus injuries
  - c. Head trauma
- 2. Postnatal trauma

b. Head trauma

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Injury to the brain parenchyma

Subarachnoid hemorrhage

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    - Hemimegalencephaly
  - b. Malformations secondary to abnormal cell migration Lissencephalies
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    - Heterotopia
  - c. Malformations secondary to abnormal late migration and cortical organization
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  - c. Lobar holoprosencephaly
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- 3. Septooptic dysplasia
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- b. Pituitary dwarfism
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- XI. Basal Cell Nevus Syndrome
- XII. Cutaneous Hemangioma-Vascular Complex Syndrome
- XIII. Chediak-Higashi Syndrome
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    - f. Hemangioblastoma
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- 1. The tools
- 2. Fetal diagnosis of hydrocephalus
- 3. Postnatal diagnosis of hydrocephalus and distortions of the brain from hydrocephalus

#### VI. Specific Categories of Hydrocephalus

- 1. Hydrocephalus resulting from excessive formation of CSF (choroid plexus papillomas)
- 2. Hydrocephalus secondary to intraventricular obstruction of CSF flow

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- 3. Hydrocephalus secondary to extraventricular obstruction of CSF
- 4. Communicating hydrocephalus
- VII. Benign Enlargement of Subarachnoid Spaces in Infants

#### VIII. Treatment of Hydrocephalus and the Resulting Complications

- 1. Radiologic assessment of third ventriculostomies
- 2. Shunt malfunctions
- 3. Shunt infections
- 4. Subdural hematoma formation
- 5. Slit ventricle syndrome
- 6. Imaging of intracranial hypotension

#### CHAPTER 9: CONGENITAL ANOMALIES OF THE SPINE

- I. Normal and Abnormal Embryogenesis of the Spine: an Overview
- II. Clinical Manifestations of Spinal Anomalies
- III. Terminology
- **IV. Imaging Techniques**

#### V. Abnormalities of Neurulation

- 1. Disorders resulting from nondisjunction: myelocele, myelomeningocele, dorsal dermal sinuses, cervical myelocystocele
- 2. Disorders resulting from premature disjunction—spinal lipomas

#### VI. Anomalies of the Caudal Cell Mass

- 1. Normal conus medullaris and filum terminale
- 2. The terminal ventricle
- 3. Fibrolipomas of the filum terminale/tight filum terminale
- 4. Syndrome of caudal regression
- 5. Terminal myelocystocele
- 6. Anterior sacral meningocele
- 7. Sacrococcygeal teratoma

#### VII. Anomalies of Development of the Notochord

- 1. The split notochord syndrome
- 2. Split cord malformation (diastematomyelia and diplomyelia)

#### VIII. Malformations of Unknown Origin

- 1. Segmental spinal dysgenesis
- 2. Dorsal meningocele
- 3. Lateral meningocele

#### IX. Congenital Tumors of the Spine

X. Syringohydromyelia

#### **CHAPTER 10: NEOPLASMS OF THE SPINE**

- II. General Imaging Characteristics of Spinal Tumors
- III. Intramedullary Tumors: Clinical Presentation, Pathology, Imaging Characteristics

#### **IV. Extramedullary Tumors**

- 1. CSF dissemination of intracranial neoplasms
- 2. Tumors of the spinal column
- 3. Meningeal tumors
- 4. Tumors of nerve roots and nerve root sheaths
- 5. Extraspinal tumors invading the epidural space

#### V. Congenital Spinal Tumors

## CHAPTER 11: INFECTIONS OF THE DEVELOPING AND MATURE NERVOUS SYSTEM

#### I. Congenital Infections

- 1. Cytomegalovirus
- 2. Toxoplasmosis
- 3. Congenital/neonatal herpes simplex encephalitis
- 4. Rubella
- 5. Congenital syphilis
- 6. Lymphocytic choriomeningitis virus
- 7. Neonatal parechovirus
- 8. Congenital varicella
- 9. Human immunodeficiency virus (perinatal transmission)
- 10. Disorders mimicking congenital infections

#### II. Meningitis and Complications

- 1. Neonatal meningitis
- 2. Meningitis in infants, children, and adolescents
- 3. Pathophysiology of meningitis
- 4. Imaging manifestations

#### III. Empyema Secondary to Sinusitis and Otomastoiditis

#### IV. Bacterial, Spirochetal, and Rickettsial Infections

- 1. Bacterial cerebritis
- 2. Brain abscess
- 3. Cat-scratch disease
- 4. Lyme disease
- 5. Rocky Mountain spotted fever

#### V. Viral Infections

- 1. General concepts
- 2. Herpesvirus family—herpes simplex, varicella zoster, Epstein-Barr, human herpesvirus-6
- 3. Nonpolio enteroviruses
- 4. Arthropod-borne viruses—eastern equine encephalitis, western equine encephalitis, Venezuelan equine encephalitis, Japanese encephalitis, St. Louis encephalitis, West Nile, dengue virus, LaCrosse encephalitis virus, California encephalitis viruses, Colorado tick fever virus
- 5. Influenza-associated encephalitis/encephalopathy
- 6. Acute cerebellitis
- 7. Rabies
- 8. Chronic viral infections—AIDS encephalopathy, progressive multifocal leukoencephalopathy, Rasmussen encephalitis, postrubella panencephalitis, subacute sclerosing encephalitis, variant Creutzfeldt-Jakob disease

#### **VI. Fungal Infections**

- 1. Neonatal candidiasis
- 2. Aspergillosis
- 3. Coccidioidosis
- 4. Cryptococcus

#### VII. Parasitic Infections

- 1. Neurocysticercosis
- 2. Visceral larva migrans
- 3. Cerebral malaria

#### VIII. Sarcoidosis

- IX. Infections of the Spine
  - 1. Discitis/osteomyelitis
  - 2. Spinal empyemas

#### X. Emerging Infections of the CNS

- 1. Nipah virus
- 2. Dengue virus
- 3. Chikungunya virus