

Contents

Preface: Introduction to Pediatric Vascular Neurosurgery **xiii**

Paul Klimo Jr, Cormac O. Maher, and Edward R. Smith

Normal and Abnormal Embryology and Development of the Intracranial Vascular System **399**

Charles Raybaud

The brain vascular system develops in such a way that it continuously adapts the supply of oxygen and other nutrients to the needs of the parenchyma. To accompany the developing brain vesicles, it evolves in several steps: superficial meningeal network first; intraventricular choroid plexuses which determine the arterial pattern; penetrating capillaries from the surface to the ventricular germinal matrix forming simple transcerebral arteriovenous loops; cortical capillaries last, mainly in the last trimester. The venous return becomes connected to both the surface and to the choroidal veins, so forming distinct meningeal and subependymal venous drainage systems, while the arteries are on the surface only. While the arterial system was determined early (week 8), the venous system is continuously remodeled by the morphological changes of the base of the skull and the expansion of the brain vesicles. Until late in gestation, the vascular system is made of simple endothelial channels in which the arterial or venous fate is determined primarily by the direction of flow.

Pediatric and Inherited Neurovascular Diseases **427**

Monique J. Vanaman, Shawn L. Hervey-Jumper, and Cormac O. Maher

Pediatric and inherited neurovascular syndromes have diverse presentations and treatments. Although many of these diseases are uncommon, they must be included in the differential diagnosis for children with strokes or hemorrhages. In neurosurgical practice, familial cavernous malformations, hereditary hemorrhagic telangiectasia (HHT), and moyamoya are the most frequently encountered of these diseases. In this article, we will discuss familial cavernomas and HHT, as well as more unusual entities such as PHACE(S) syndrome, Klippel-Trenaunay syndrome, Wyburn-Mason syndrome, sinus pericranii, radiation-induced vasculopathy, and blue rubber bleb nevus (BRBN) syndrome. Moyamoya disease is covered in several other articles in this volume.

Diagnosis and Management of Arteriovenous Malformations in Children **443**

Toba N. Niazi, Paul Klimo Jr, Richard C.E. Anderson, and Corey Raffel

Arteriovenous malformation (AVM) is the most common cause of spontaneous intraparenchymal hemorrhage in children, excluding hemorrhages of prematurity and early infancy. Because most children diagnosed with an AVM undergo initial treatment emergently, the natural history of AVMs in the pediatric population is not well understood. Most pediatric AVMs do not come to clinical attention unless they hemorrhage. Therefore, their optimal management remains controversial. Children with intracranial AVMs represent a special challenge in that they harbor unacceptable lifelong risks of hemorrhage and potential neurologic deficits. Patients should be evaluated on a case-by-case basis to determine the best multidisciplinary treatment regimen that can be used to preserve neurologic function and eradicate

the AVM with the lowest risk of mortality. Successful treatment depends on the location and size of the AVM, its hemodynamic properties, the clinical condition of the patient, and the treatment modality selected. The armamentarium for AVM management has grown with technological advances and now includes microsurgical resection, endovascular embolization, radiosurgery, or any combination of these modalities. Microsurgical resection remains the gold standard for treatment of accessible pediatric AVMs, especially in cases that present with intracranial hemorrhage. Newer modalities, such as embolization and radiosurgery, have provided additional tools to help children with large or deep-seated lesions that would be deemed unresectable with microsurgical techniques alone. Long-term follow-up with repeated diagnostic imaging is important despite complete obliteration of the lesion to rule out the small possibility of AVM recurrence.

Stereotactic Radiosurgery for Pediatric Arteriovenous Malformations

457

Andrew B. Foy, Nicholas Wetjen, and Bruce E. Pollock

Children with intracranial arteriovenous malformations (AVM) have a high cumulative risk of hemorrhage and therefore effective treatment of AVMs in the pediatric population is imperative. Treatment options include microsurgical resection, endovascular embolization, staged or single fraction radiosurgery, or some combination of these treatments, with the ultimate goal of eliminating the risk of hemorrhage. In this article the authors review the current data on the use of radiosurgery for the treatment of childhood AVMs. Factors associated with successful AVM radiosurgery in this population are examined, and comparisons with outcomes in adult patients are reviewed.

Classification and Endovascular Management of Pediatric Cerebral Vascular Malformations

463

Timo Krings, Sasikhan Geibprasert, and Karel terBrugge

Pediatric vascular malformations of the central nervous system differ from those seen in adults. Their classification may be based on symptoms, pathomechanics, patient's age, morphologic features, or presumed etiology. This review describes the different classification schemes and the endovascular management options of these rare and challenging diseases. The proposed etiologic classification of pediatric vascular malformations may add to our understanding of these diseases in general because the phenotypic expression of a given vascular malformation can shed light on the nature and timing of the causative agent, thereby potentially opening up treatment modalities in the future that are directed against the triggering event rather than against the clinical manifestations or the morphologic appearance. With current endovascular methods, most vascular diseases can be approached safely and with good clinical results.

Cavernous Malformations

483

Edward R. Smith and R. Michael Scott

Cavernous malformations (CMs) are vascular lesions found in the central nervous system (CNS) and throughout the body and have been called cavernomas, cavernous angiomas, and cavernous hemangiomas. This article discusses the epidemiology, natural history, diagnosis, treatment and follow-up of children who are found to harbor these lesions. CMs affect children by causing hemorrhage, seizure, focal neurologic deficits, and headache. Diagnosis is best made with magnetic resonance imaging. Patients with multiple lesions should be referred for genetic evaluation and counseling. Individuals with symptomatic, growing, or hemorrhagic malformations

should be considered for surgical resection. Close follow-up after diagnosis and treatment is helpful to identify lesion progression or recurrence.

Pediatric Intracranial Aneurysms

491

Brian J. Jian, Steven W. Hetts, Michael T. Lawton, and Nalin Gupta

Intracranial pediatric aneurysms arising in children are rare. The treatment of these lesions requires both an understanding of their unique features as well as surgical, interventional, and pediatric critical care expertise offered through a multidisciplinary setting. The patient population, clinical presentation, complications, and trends in treatments are discussed in this article.

Spinal Cord Vascular Malformations in Children

503

Debbie Song, Hugh J.L. Garton, Daniel K. Fahim, and Cormac O. Maher

Spinal vascular malformations comprise a diverse group of abnormalities, including arteriovenous malformations (AVMs), cavernous malformations, dural arteriovenous fistulas (AVFs), and capillary telangiectasias. These conditions each have distinct causes, presentations, radiologic appearances, and natural histories. This article explores the presentation, natural history, investigation, and treatment of spinal AVMs, spinal AVFs, and spinal cavernous malformations.

Cerebral Venous Sinus (Sinovenous) Thrombosis in Children

511

Nomazulu Dlamini, Lori Billingham, and Fenella J. Kirkham

Cerebral venous sinus (sinovenous) thrombosis (CSVT) in childhood is a rare, but underrecognized, disorder, typically of multifactorial etiology, with neurologic sequelae apparent in up to 40% of survivors and mortality approaching 10%. There is an expanding spectrum of perinatal brain injury associated with neonatal CSVT. Although there is considerable overlap in risk factors for CSVT in neonates and older infants and children, specific differences exist between the groups. Clinical symptoms are frequently nonspecific, which may obscure the diagnosis and delay treatment. While morbidity and mortality are significant, CSVT recurs less commonly than arterial ischemic stroke in children. Appropriate management may reduce the risk of recurrence and improve outcome, however there are no randomized controlled trials to support the use of anticoagulation in children. Although commonly employed in many centers, this practice remains controversial, highlighting the continued need for high-quality studies. This article reviews the literature pertaining to pediatric venous sinus thrombosis.

Traumatic Intracranial and Extracranial Vascular Injuries in Children

529

Roukoz B. Chamoun and Andrew Jeal

Trauma continues to be the leading cause of death in children older than 1 year of age. Although vascular injuries are uncommon, they contribute significantly to the mortality and morbidity related to traumatic injuries in the pediatric age group. In a recently reported large series of children, the head and neck location constituted 19.4% of all pediatric vascular injuries and accounted for most of the mortality observed. Catheter angiography is still considered as the gold standard diagnostic modality. However, because of its invasive nature, other techniques such as computed tomography angiography and magnetic resonance angiography are emerging as alternative diagnostic screening tools. Traumatic vascular injuries can involve the

carotid as well as the vertebral arteries. They can be extracranial or intracranial. As a result, traumatic vascular injuries are a heterogeneous group of entities with potential significant implication on the natural history and prognosis. The optimal management of these injuries remains unclear and current practice is largely individualized. This report reviews the available literature regarding the current trends in diagnosis and management of pediatric traumatic vascular injuries.

Moyamoya: Epidemiology, Presentation, and Diagnosis **543**

Edward R. Smith and R. Michael Scott

Moyamoya syndrome is an increasingly recognized arteriopathy associated with cerebral ischemia and has been associated with approximately 6% of childhood strokes. It is characterized by chronic progressive stenosis at the apices of the intracranial internal carotid arteries, including the proximal anterior cerebral arteries and middle cerebral arteries and ultimately results in decreased cerebral blood flow with an increased resultant risk of stroke. This article discusses the epidemiology, presentation, and diagnosis of this condition in children.

Indirect Revascularization Techniques for Treating Moyamoya Disease **553**

Neil N. Patel, Francesco T. Mangano, and Paul Klimo Jr

There have been many indirect revascularization techniques described by surgeons for the treatment of moyamoya disease. These surgical procedures are typically used more commonly in pediatric, than in adults', cases. Some of the techniques include: cervical sympathectomy, omental transplantation, multiple burr holes, encephalo-myo-synangiosis (EMS), encephalo-arterio-synangiosis (EAS), encephalo-duro-synangiosis (EDS), encephalo-myo-arterio-synangiosis (EMAS), encephalo-duro-arterio-synangiosis (EDAS), encephalo-duro-arterio-myo-synangiosis (EDAMS), encephalo-duro-galeo (periosteal)-synangiosis (EDGS), and combinations of all the above. This chapter will detail the technical aspects of many of these procedures and some of the reported clinical outcomes.

Direct Bypass Techniques for the Treatment of Pediatric Moyamoya Disease **565**

Raphael Guzman and Gary Steinberg

Moyamoya is an increasingly recognized cause of stroke in children and adults. Identification of the disease early in its course with prompt institution of therapy is critical to providing the best outcome for patients. Revascularization surgery seems to be effective in preventing stroke in moyamoya, with direct techniques providing durable protection when performed at experienced centers.

Index **575**