Common Neurosurgical Conditions in the Pediatric Practice

Recognition and Management

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Preface

Within medicine, increasing subspecialization has created an almost impossible task; primary care providers are now asked to triage their patients and provide appropriate referrals within a dizzying array of diverse medical fields. Pediatric neurosurgery remains one of a number of highly specialized fields within which most pediatric practitioners including physicians, physician's assistants, and nurse practitioners have never received any training. This fact, combined with the rarity of pediatric neurosurgical diseases, the acuity with which many of them present, the need for rapid triage and tertiary care, and parental anxiety when receiving a neurologic diagnosis, creates a challenging situation for providers caring for children with these conditions.

Several years ago, we hosted a CME event in New York City bearing the same name as this textbook. We solicited comments from attendees and were pleased that most participants felt we were filling a real deficiency in medical education—the cross-fertilization between subspecialties. In fact, a recent survey of pediatricians through an AAP initiative supports our suspicion that many primary care providers felt uncomfortable managing patients with neurosurgical conditions. This survey also suggested a number of neurosurgical topics that pediatricians felt they wanted more assistance in managing and triaging. All of these topics, and many others, are covered from both a neurosurgical and pediatric perspective in this textbook.

In fact, this textbook was designed around a central concept:

> Pediatricians and subspecialists need to have a closer working relationship to make childhood medicine work seamlessly in 2016 and beyond.

A pediatrician in a busy practice should not be expected to explain to a concerned parent what the differences in treatment options may be between different types of brain tumors; however all pediatric practitioners should be able to discern between benign and worrisome headaches and to decide when imaging, referral to neurology, or triage to an emergency department is warranted. The relationship between primary care physicians and subspecialists such as pediatric neurosurgeons is obviously at the crux of this book, but should also be at the forefront of mutual efforts to improve care of children. This textbook will never be able to replace the trust and confidence that is built through years of communication and shared patient experiences.

To highlight our belief in the strength of the relationship between primary care providers and subspecialists, this book was envisioned by and co-edited by a pediatric neurosurgeon and a pediatrician each providing the perspective

from within the lens through which they view these common neurosurgical conditions. We hope the vignettes provide real-life flavor to experiences many of you have had, or will have. We expect that the "red flags," "pediatrician's perspective," and references will serve as focused reviews on many of these topics. When reading a newly arrived imaging report, or caring for a child with an unfamiliar diagnosis, these chapters can serve as a medical refresher before calling a parent back with unexpected news or reviewing a finding in the office.

The book is organized into parts that loosely approximate the neurologic development of a child and address issues that are commonly encountered. The first part reviews neurologic development and birth-related trauma commonly seen in the neonatal intensive care unit including issues such as brain injury, brachial plexus injury, and hematomas. The second part addresses findings commonly encountered in the first month of life in the pediatrician's office. Lumps and bumps, manifestations of neurocutaneous diseases, or tethered cord stigmata are all reviewed in depth. The third part is a comprehensive review of hydrocephalus from birth-related intraventricular hemorrhage through the work-up of macrocephaly and headache management. Part 4 is an important group of chapters describing up to the date thoughts on imaging the central nervous system in children from prenatal ultrasound through MRI and CT including when these different modalities are important to select and the risks associated with each. The fifth grouping of chapters consists of explorations of common neurosurgical conditions that many pediatricians are uncomfortable dealing with, including brain tumors, spasticity, and vascular lesions to use as a reference tool when caring for a complex neurosurgical patient. Finally a series of chapters related to head trauma concludes the textbook, including sections on non-accidental trauma and concussion management.

Most importantly, we hope the topics we cover in the ensuing chapters provide you with some measure of confidence in dealing with conditions you may feel less comfortable with than others, and provide a framework within which you can direct subspecialized care.

We would very much like to take this opportunity to thank our mentors within medicine—Dan Cohen, Gary Edelstein, Sarah Long, Phil Stieg, and Mark Souweidane—for supporting and inspiring our careers and providing continual support and guidance through the years. Educating subsequent generations of pediatricians and neurosurgeons with the lessons you taught us is your greatest legacy and we are proud to be within your lineages.

To our parents and families who instilled within us the belief that education and academics can be at the center of a rich and meaningful life, thank you for supporting our endless training and sharing pride in our demanding but fulfilling careers. Everything we have learned began with your confidence in us and now allows us to begin to pass this on to the next generation of pediatric physicians.

Finally, we are forever indebted to Richard Lansing and Joni Fraser from Springer for seemingly unending patience as we tried to balance our busy medical careers and four young boys at home with an innocent foray into the editing and publishing world. We hope your confidence in us is rewarded in

the impact this book will have and the children benefiting from the shared knowledge going forward.

To our readers, best of luck caring for your patients with these neurologic conditions—there is often no greater challenge in medicine but also often no greater reward!

New York, NY, USA

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Part I

Development of the Brain and Spine

Normal Development of the Skull and Brain

1

Waleed A. Azab

Introduction

The cranial vault or neurocranium encloses the brain, meninges, and cerebrospinal fluid. *The single most important stimulus for head growth during infancy and childhood is brain growth*, and this will be a recurrent theme when trying to consider the driving forces behind many neurosurgical or central nervous system-related findings in your young patients. In the subsequent chapters, we will systematically introduce some pathologic conditions pertaining to the intersection of brain and skull development throughout adolescence. The abnormally large head, small head, and misshapen heads are all of concern to parents and medical care providers alike. In this chapter we will introduce a framework of thinking to use as a reference tool when evaluating your patients. In the subsequent chapter, we will provide a simple and yet comprehensive framework for the pediatric neurologic examination and then begin our journey through the various manifestations of normal, gone awry.

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In the newborn child, bones of the cranial vault are separated by intervening sutures. At points of multiple bony oppositions, sutures widen and assume the shape of fontanels. Sutures and fontanels close at quite wide age ranges (discussed more in Chap. 6). Sutures facilitate deformation of the head during delivery and allow uniform expansion of the calvarium during brain growth. The "cone-shaped" head that so many infants manifest after vaginal delivery should be an opportunity to tell anxious parents how well evolution and anatomy have conspired to allow babies with such large brains to develop intrauterinely for so long! In an otherwise normal (nonsyndromic) child, and the absence of a midsagittal ridge (Chap. 6), birth-related cranial deformity should not be cause for concern. Abnormal head shapes can of course be caused by craniosynostosis or deformational plagiocephaly. Premature closure of cranial sutures results in craniosynostosis, a condition that usually needs surgical treatment. Deformational (positional) plagiocephaly refers to the deformation of the head characterized by a persistent flattening on the side with an asymmetric head shape and misalignment of the ears. Plagiocephaly is a benign condition that should be differentiated from craniosynostosis and is usually treated conservatively. These conditions are very easily differentiated from the normal skull and are discussed in detail later.

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Normal Skull Development

Enclosed within the skull bones, brain growth continues exponentially in the first 2 years of life. Postnatal growth of the skull is characterized by changing proportions of its components. In addition to the continued growth of the frontal and parietal bones, the squamous temporal bone increases in size so that it contributes a greater proportion of the skull vault in the adult than in the neonate [1]. The skull without the mandible is called the cranium (Fig. 1.1), and may be subdivided into two regions. The cranial vault or neurocranium encloses the brain, meninges, and cerebrospinal fluid, while the facial skeleton or viscerocranium hangs down from the front of the neurocranium and encloses the organs of special sense [2]. At birth, the skull is proportionally large, reflecting the importance of brain maturation. The volume of the neurocranium compared to that of the face is about 8:1 in the newly born infant. This ratio becomes 5:1 by 2 years, 3:1 at 6 years, and 2:1 in the adult [3]. The major part of the skull vault is formed by paired frontal and parietal bones and a single interparietal bone. The squamous part of the temporal bones and the membranous part of the greater wing of sphenoid contribute to the lateral walls of the skull [2].

The bones of the skull base are formed by endochondral ossification [1]. Bone formation of the skull vault takes place through a process of intramembranous ossification from a mesenchymal layer situated between the dermal mesenchyme and the meningeal membranes [4]. The mesenchymal cells condensate then differentiate into osteoblasts and deposit extracellular matrix. Ossification proceeds radially from these condensations to ultimately form the bones of the skull vault [5].

In the newborn, the membranous bones of the vault are separated by the intervening sutures. At the points of intersection, sutures widen and assume the shape of fontanels. The larger anterior fontanel is at the intersection of the sagittal, coronal, and metopic sutures, and the posterior fontanel is at the intersection of the sagittal and lambdoid sutures (Fig. 1.2). The most significant growth of the skull occurs along the sagittal and coronal sutures [6]. Sutures facilitate deformation of the head during delivery and allow uniform expansion of the calvarium during brain

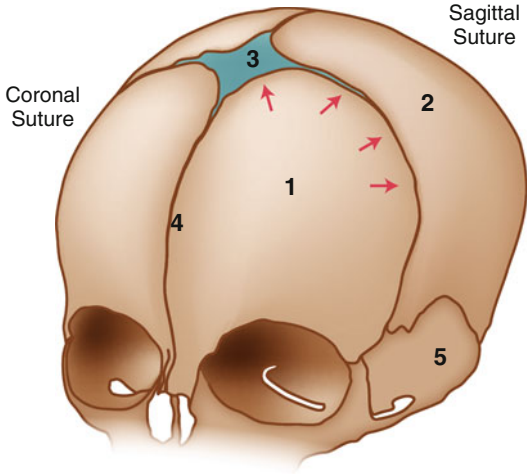

Fig. 1.1 Neonatal skull. Edge of the frontal bank of left coronal suture is marked with *arrows*. (*1*) Frontal bone, (*2*) parietal bone, (*3*) anterior fontanel, (*4*) metopic suture, (*5*) squamous temporal bone

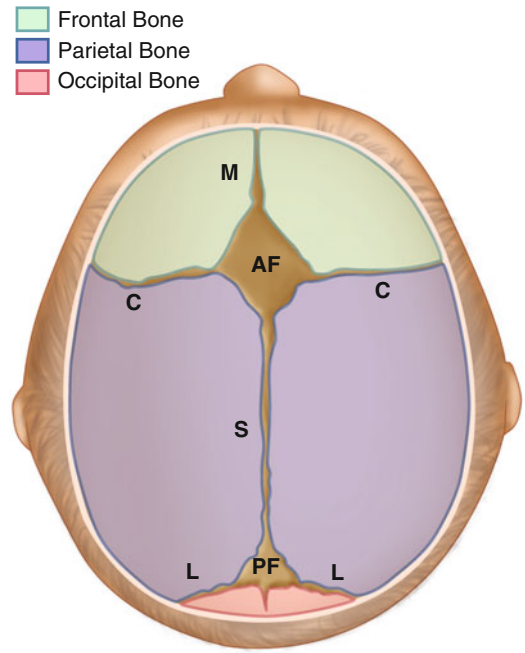

Fig. 1.2 Illustration of the vault of the skull in an infant viewed from above. *AF* anterior fontanel, *C* coronal suture, *L* lambdoid suture, *M* metopic suture, *PF* posterior fontanel, *S* sagittal suture

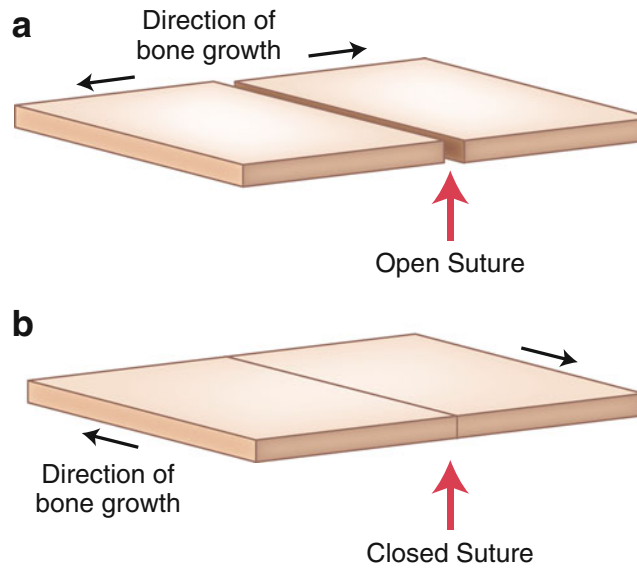

Fig. 1.3 Virchow's law

growth by its fibrous connective tissue content [7, 8]. Sutures regulate the balance between proliferation and differentiation of the osteogenic precursors through multiple molecular pathways and are the principal growth centers in the skull for the first few years of life [5, 9]. As a rule, the growth of the skull is perpendicular to the open suture lines and parallel to a fused suture (Virchow's law) (Fig. 1.3) [7]. Closure of the various fontanels and sutures takes place at specific age ranges, but it is rarely any cause for concern, in isolation, in a developmentally normal child with an otherwise normal cranial morphology when a fontanel closes earlier or later than expected (Table 1.1).

Table 1.1 Normal age ranges of closure for fontanels and cranial sutures

Fontanel or suture	Age of closure
• Anterior fontanel	12–18 months
• Posterior fontanel	3–6 months
• Posterolateral fontanel	24 months
• Anterolateral fontanel	3 months
• Metopic suture	3–8 months
• Coronal sutures	~35 years
• Lambdoid sutures	~35 years
• Sagittal suture	~35 years
• Squamosal sutures	~35 years

Modified from Pekçevik Y, Hasbay E, Pekçevik R. Three-dimensional CT imaging in pediatric calvarial pathologies. Diagn Interv Radiol. 2013 Nov–Dec;19(6):488–94

The base of the infant skull, on the other hand, contains multiple cartilaginous joints or synchondroses located between the sphenoid and ethmoid bones anteriorly and between the sphenoid and occipital bones posteriorly. Growth of the skull base and consequent cranial lengthening is largely independent of cerebral growth and takes place mostly at the synchondroses between the sphenoid and occipital bones. In the sphenoid region, three prominent synchondroses can be identified and are named the fronto-sphenoid, the inter-sphenoid, and the spheno-occipital. The first two usually close by age 2, but the spheno-occipital synchondrosis may be visible on lateral radiographs of the skull base until the age of 18 years [2, 10].

The single most important stimulus for head growth during infancy and childhood is brain growth [11]. Throughout the period of rapid development of the brain, pressure is exerted on the inner table of the skull, which accommodates to the increasing size of the brain. Such an adaptation is facilitated by the membranous fontanels, which remain open until maximal brain growth has been attained. Accurate assessment of head growth is therefore one of the most important aspects of the neurologic examination of infants and young children [11].

Normal Development of the Brain

The development of the nervous system occurs through the interaction of several synchronized processes, some of which are complete before birth, while others continue into adulthood [12]. The central nervous system begins to develop in the human fetus 2–3 weeks after fertilization of the oocyte. From 4 to 12 weeks of gestation, ectodermal tissues of the neural tube begin to differentiate into the precursors of the various structures of the nervous system. The forebrain and facial structures develop at one end and the spinal cord at the other. The hollow center of the tube in the region of the future brain eventually develops into the ventricles. Regions called proliferative or ventricular zones form in the vicinity of the ventricles and differentiate into the site for the division and origin of cortical and subcortical neurons [12, 13]. Between weeks 12 and 20 of gestation, neurons migrate from the ventricular and adjacent subventricular zones along a scaffolding of glial cells toward their determined final destinations in the cortex [12, 14].

Subsequently, a period of rapid programmed cell death occurs, reducing neuronal populations by half between 24 weeks of gestation and 4 weeks after birth. The cell bodies of the neurons are primarily found in the gray matter of the brain. Their myelinated axons form white matter. Myelination begins at the brain stem by 29 weeks and generally proceeds from inferior to superior and posterior to anterior. Proximal pathways tend to myelinate before distal, sensory before motor, and projection before association. Although most major tracts are significantly myelinated by early childhood, some axons continue to myelinate into the second and third decades of life [12].

Another major developmental process is the proliferation and organization of synapses, which begins around the 20th week of gestation [12]. The rate of synapse formation peaks after the 34th week of gestation reaching about 40,000 new synapses per second [13]. A rapid increase in synaptic density occurs after birth with a number estimate at the age of 2 years that is around 50 % greater than the typical number in adults [15]. Due to increasing cell and synaptic density, beginning at approximately 15 weeks of gestation, the surface of the growing brain begins to fold into sulci and gyri. The major sulci, except for the occipital lobe, are in place by 28 weeks of gestation, after which secondary and tertiary sulci are elaborated, with nearly all gyri present by birth. The sulcal and gyral patterns continue to increase in complexity after birth [12]. Rapid brain growth takes place in the first 2 years of life reaching 80–95 % of its adult size [16].

Abnormal Head Size

Evaluation of the Head Circumference

In 1968, Nellhaus compiled graphs of head circumference in children of both sexes from birth to 18 years of age [17]. Detailed tables and percentile charts based on measurements taken from a very large number of subjects have become available through the WHO Multicentre Growth Reference Study Group which established the WHO Child Growth Standards for head circumference for age. We have included these charts for easy accessibility inside the cover of this book. Detailed normative data and percentile charts for boys and girls, specific charts that account for prematurity, and specific chart for children with certain specific etiologies such as Down's syndrome can be accessed online at the WHO website (http://www.who.int/childgrowth/standards/hc_for_age/en/) [18]. A head circumference that is two standard deviations above or below the mean for age requires investigation and explanation [11].

Macrocephaly

Macrocephaly is defined as a head circumference more than two standard deviations for age and sex above the mean. It can be caused by various benign or pathological conditions (Table 1.2). Common causes of macrocephaly include familial megalencephaly (larger-than-normal brain mass), benign extracerebral collections of infancy (BECC), and hydrocephalus [21], the workup and treatment of which will be discussed in more detail in Chaps. 13 and 14.

Table 1.2 Causes of macrocephaly [11, 19–21]

• Familial megalencephaly (larger-than-normal brain mass)
• Benign extracerebral collections of infancy (BECC)
• Hydrocephalus
• Hydranencephaly
• Brain tumors
• Intracranial cysts
• Pseudotumor cerebri
• Subdural hematomas or hygroma
• Rebound or "catch-up" brain growth (after prematurity or serious illness)
• Genetic, metabolic, and dysplastic syndromes (e.g., neurofibromatosis, Soto syndrome, mucopolysaccharidoses, hemimegalencephaly, achondroplasia)
• Lipid storage disease, leukodystrophies, cranial dysplasias, and marrow hyperplasia (chronic hemolytic anemias)

Table 1.3 Causes of microcephaly [3, 11]

• Trisomies 13, 18, 21
• Lissencephaly, schizencephaly
• Rubenstein-Taybi, Cornelia de Lange, Angelman syndromes
• Fetal alcohol syndrome, anticonvulsants, maternal phenylketonuria (PKU)
• Intrauterine infections; TORCHS
• Radiation in the first and second trimesters
• Placental insufficiency
• Autosomal dominant and autosomal recessive familial microcephaly
• Hypoxic-ischemic injury, birth asphyxia
• Bacterial meningitis (especially group B streptococci), viral encephalitis (enterovirus, herpes simplex)
• Glut-1 deficiency, PKU, and maple syrup urine disease
• Tay-Sachs and Krabbe's diseases

Detailed history, neurological examination (especially for signs of high intracranial pressure), evaluation of developmental milestones, and assessment of head growth rate through serial head circumference measurements are important for differential diagnosis, urgency of imaging, and further management [19]. Macrocephaly with normal growth rate and normal neurological examination is reassuring and is characteristic of benign megalencephaly which is usually familial [21]. Rapid head growth rate with loss of developmental milestones or other neurologic findings, on the contrary, suggests an increased intracranial pressure, often caused by hydrocephalus or neoplasms [11].

Macrocephaly and accelerated head growth with normal neurological exam and absence of evidence of elevated intracranial pressure may occur as nonprogressive subarachnoid space dilatation with or without ventricular enlargement. This pattern is most commonly referred to as benign extracerebral collection of infancy (BECC). The cause is unknown, but it may be related to delayed development of parasagittal dural channels responsible for cerebrospinal fluid (CSF) resorption in young children (who have few arachnoid villi). Accelerated head growth may continue until 12–18 months of age and then usually stabilizes as a form of megalencephaly. Imaging features of BECC include normal to mildly enlarged lateral and third ventricles and symmetric enlargement of the frontal subarachnoid spaces, interhemispheric fissure, and Sylvian fissures [21].

Microcephaly

Microcephaly has fewer neurosurgical implications than macrocephaly. It is defined as a head circumference more than two standard deviations below the mean for age and sex which is by definition microcephaly [11]. Some causes of microcephaly are listed in Table 1.3 and has an expanded discussion dedicated to the topic within Chap. 6.

Abnormal Head Shape

Craniosynostosis

A detailed review of the various craniosynostoses and their management is beyond the scope of this chapter; Chap. 6 is dedicated entirely to this topic; however, a brief account on some features of this entity is presented here. Premature closure of cranial sutures results in craniosynostosis [22]. The incidence of craniosynostosis is approximately 1 in 2500 live births, and the condition is broadly classified into non-syndromic and syndromic craniosynostosis [23–25]. Unlike the syndromic

type, non-syndromic synostosis is not associated with other dysmorphisms of the face, trunk, or extremities. Furthermore, a non-syndromic craniosynostosis typically involves a single suture [26]. The most commonly affected sutures in descending order are the sagittal suture, followed by the unilateral coronal, bilateral coronal, metopic, and lambdoid sutures. Syndromic craniosynostosis is much less common and appears to be a more generalized disorder of mesenchymal development [25].

Sagittal synostosis is caused by fusion of the sagittal suture and results in a boat-shaped deformity of the skull or scaphocephaly, with growth restriction in width and compensatory excessive growth in calvarial length in the anteroposterior direction. This growth pattern leads to varying degrees of frontal bossing and occipital coning [26]. *Unicoronal synostosis* results in anterior plagiocephaly, with ipsilateral flattening of the forehead on the affected side and contralateral bulging of the frontoparietal calvaria. The growth restriction of the forehead in unicoronal synostosis results in a facial twist. It is believed that the compensatory pressure of the ipsilateral temporal lobe pushes the maxilla forward leading to forward displacement of the ipsilateral zygoma and rotation of the maxilla so that the nasal tip is deviated to the contralateral side. Bilateral coronal fusion produces brachycephaly or skull shortening in the anteroposterior diameter and skull lengthening in a cranio-caudal direction or turricephaly [27]. *Metopic synostosis* results in trigonocephaly or a triangular-shaped forehead with bifrontal and bitemporal narrowing and parietal and occipital prominence. Additionally, hypotelorism and a low nasal dorsum with epicanthal folds are present [25]. *Unilateral lambdoid synostosis* is characterized by abnormal shape of the occipital region and an ipsilateral mastoid bulge; the affected lambdoid suture is thickened and ridging with ipsilateral downward tilting of the occipital skull base. The external auditory canal and entire ear is displaced inferiorly, a clinical finding that is more reliable than inspecting for anterior or posterior displacement of the ear in differentiating positional plagiocephaly and lambdoidal synostosis. A compensatory bulge of the contralateral posterior parietal region gives the skull an oblique towering appearance when seen from the back [28–36].

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The Neurologic Exam in Neonates and Toddlers

2

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Introduction

Performing a comprehensive neurological examination in children is sometimes considered a challenge by non-neurologists. A neurologic exam which tests all aspects of all neurologic modalities can quite literally take several hours to perform. On the other hand, a very good, thorough neurologic exam which yields substantial relevant information can be performed in under 5 min. Adherence to a systematic framework or approach to the examination, appropriate for the age and abilities of the child, can be extremely helpful in simplifying the basic questions: Is this child normal, and if not, why not, and how do I describe it? This chapter will focus on how to perform some of the routine elements of the neurologic examination with tips on how to tailor the exam for various age groups.

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The History

As always, obtaining a solid history of present illness is essential for directing the physical exam and establishing a diagnosis, and one is often able to narrow the differential diagnosis substantially based on history alone. As most people who work with children have already learned, simple observation of the child while obtaining the history can also influence one's approach toward obtaining information.

Birth History

It is likely not necessary to remind anyone with a background in pediatrics that any history in children should include details of the pregnancy and birth. Many neurologic disorders, whether genetic or acquired, can begin in pregnancy. Complications in pregnancy, such as growth restriction, failure to progress, fetal distress, or prolonged labor, may indicate antenatal disorders rather than any process which began as a result of the actual birth process itself. One should evaluate for maternal antenatal factors such as illnesses and teratogenic exposures. Of course, the gestational age at which the child was born will also help establish a developmentally appropriate norm. If known, Apgar scores may also be informative and give a general clue as to the onset or timing of childhood disorders.

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