Child with Acute Neurological Emergency

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Abstract

Children with acute neurological emergencies present to the ER with a wide spectrum of symptoms and signs. Neuroimaging plays an important role because of limitations such as gathering an accurate patient history and difficulties in performing a detailed neurological examination in the ER, particularly in young patients. The goal of this chapter is to discuss the neuroimaging findings of the most frequent causes of acute emergencies in children, as well as of some less frequently encountered entities.

Keywords

Neurological emergencies · Stroke · Hemorrhages
Infections · Trauma · Seizures

Learning Objectives

- To understand the common acute neurological disorders in children encountered in the emergency room.
- To learn basic neuroimaging findings of acute pediatric neurological emergencies.

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14.1 Introduction

Neurological emergencies account for about one-third of the highest severity codes encountered in the pediatric emergency department [1]. Children present to the emergency room (ER) with a wide range of symptoms, including headache, fever, nausea, vomiting, altered mental status, coma, and physical signs of trauma. Neuroimaging plays an important role because of limitations such as gathering an accurate patient history and difficulties in performing a detailed neurological examination in the ER, particularly in young patients. When faced with a child with an acute neurological emergency, radiologists have to (1) decide whether neuroimaging is required emergently, (2) choose the most appropriate imaging modality based on the institutional availability and the clinical status of the patient, and (3) consider specific issues related to the pediatric age (e.g., higher sensitivity to radiation compared to adults, need for sedation or general anesthesia, different appearance of the pediatric brain related to the age of maturation and development) [2].

Computed tomography (CT) is most often the initial neuroimaging modality of choice in acutely ill children. This method is widely available and fast, and with modern multidetector CT scanners, it is possible to acquire submillimeter-thin cross-sectional images that can be used to render multiplanar reformats and three-dimensional (3D) images. This allows rapid detection of skull and facial fractures. However, the primary disadvantage of CT is that it requires ionizing radiation [2].

Magnetic resonance imaging (MRI) overcomes CT's ionizing radiation limitation and also allows superior anatomic details and various tissue contrast (especially for the evaluation of the posterior fossa). It may also give more accurate information about the timing and quality of injury. Furthermore, MRI can provide multiple advanced and functional sequences progressively incorporated into acute pediatric neuroimaging protocols [2]. These advantages make MRI the preferred diagnostic modality in children with acute

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neurological emergencies. Advanced MRI techniques include susceptibility-weighted imaging (SWI), diffusion-weighted imaging (DWI), diffusion-tensor imaging (DTI), magnetic resonance spectroscopy (MRS), and perfusion-weighted imaging (PWI) including arterial spin labeling (ASL). MRI's main disadvantage is that it generally requires longer scan times, but ultrafast MRI protocols may be considered instead of CT in a time-sensitive setting [3].

Lastly, among the challenges radiologists face is the consideration that children are not small adults and pediatric neuroimaging differs from adults [4]. When evaluating brain MRIs of children, radiologists should be familiar with normal variants, congenital or developmental disorders, and age-dependent changes to provide reliable differential diagnosis and reduce morbidity and mortality.

This chapter aims to discuss the neuroimaging findings of the most frequent causes of acute emergencies in children as well as of some less frequently encountered entities.

14.2 Stroke

Arterial ischemic stroke (AIS) is considered a rare entity in the pediatric population. However, a lack of familiarity often results in delayed diagnosis and high morbidity and mortality rates. Compared to adults, stroke in children has different risk factors and clinical presentations that affect the diagnostic approach, management, and outcome. The most relevant risk factors include (1) cardiac causes (e.g., congenital heart defects, endocarditis, cardiomyopathies, valvulopathies), (2) hematological diseases (e.g., sickle cell disease, coagulopathies), (3) vascular causes (e.g., arteriovenous malformations (AVM), arteriopathies), and (4) genetic/hereditary conditions (e.g., neurofibromatosis type I, Fabry disease) [5]. Acute ischemic strokes presenting in the perinatal period are a specific subset of vascular injury in term newborns and, while they are characterized by different etiologies than in older age groups (i.e., placental emboli, coagulopathies), are often eventually labeled as idiopathic [6].

Acute infarctions may not be visible on ultrasound (US) and CT within the first 24 h of the event. In contrast, MRI provides high sensitivity in detecting acute stroke in children and is considered the preferred imaging modality. DWI and ADC mapping sequences are pivotal. Arterial infarcts appear hyperintense on DWI and hypointense on ADC maps within minutes of occurrence (Fig. 14.1a, b). T2*gradient echo (GRE) and SWI are the most sensitive in detecting intracranial blood and should also be included. Other recommended MRI sequences include T1- and T2-weighted images (WI) to assess myelination, intra- or extra-axial products, and edema [7].

Furthermore, combined analysis of DWI/PWI findings can identify potentially salvageable brain tissue, i.e., "ischemic penumbra." The ischemic penumbra appears hypoperfused but does not reveal a matching diffusion restriction (DWI/PWI mismatch), unlike the ischemic core which shows matching areas of hypoperfusion and restricted diffusion (Fig. 14.1c) [8].

In addition, MRA of the brain should be performed to assess vascular supply, stenosis, or occlusion (Fig. 14.1d).

Hemorrhagic venous stroke (VS) in the neonatal period may occur as a cerebral venous sinus thrombosis (CVST) complication. Multiple risk factors include maternal and fetal causes (e.g., gestational diabetes, preeclampsia, neonatal sepsis, dehydration, prothrombotic state) [8]. Imaging findings may be misleading and include (1) intraventricular hemorrhage in a term neonate, (2) thalamic, caudate, or parietal hemorrhage, and (3) ischemia not respecting an arterial distribution [7]. MRI remains the preferred modality to diagnose CVST. The appearance of venous thrombus on T1- and T2-WI depends on the age of the clot. T2*GRE and SWI are both indicated to detect venous stasis. Along with MRI, magnetic resonance venography (MRV) should be considered in the setting of an "unexplained" spontaneous hemorrhage (Fig. 14.2) [9].

Numerous other neurologic conditions may clinically mimic pediatric stroke (e.g., hypoglycemia, hemiplegic migraine, seizure, postictal paresis, infection) in up to 20% of children with the initial suspicion of acute stroke [9, 10]. The high frequency underlines the value of MRI in evaluating suspected acute stroke.

Key Points

- Neuroimaging is extremely time sensitive in children with suspected ischemic or hemorrhagic stroke.
- MRI with DWI/DTI and ADC sequences is the established modality of choice for accurate diagnosis.

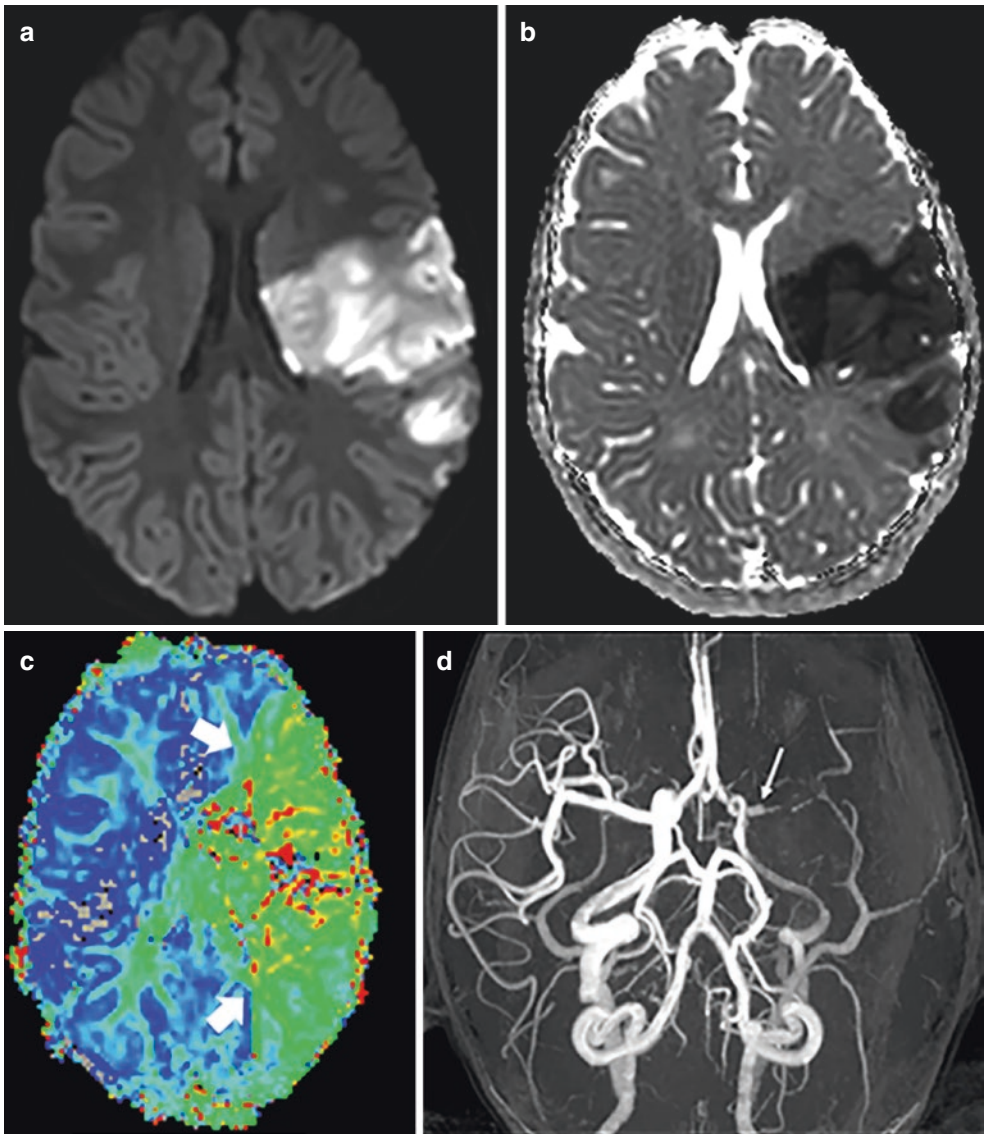

Fig. 14.1 Acute ischemic stroke due to focal cerebral arteriopathy in a 7-year-old girl. (**a**) Diffusion-weighted image and (**b**) corresponding axial ADC map show large area of restricted diffusivity in the left cerebral hemisphere, consistent with ischemic core in the territory of the left middle cerebral artery (MCA). (**c**) Perfusion-weighted image (dynamic susceptibility contrast T2*), mean transit time map colorized map shows larger area of reduced perfusion (arrows) corresponding to the totality of the left MCA territory and indicating that a large ischemic penumbra of potentially salvageable nervous tissues exists. (**d**) 3D time-of-flight arterial MR angiogram, maximum intensity projection reveals almost complete obstruction of the M1 segment of the left MCA

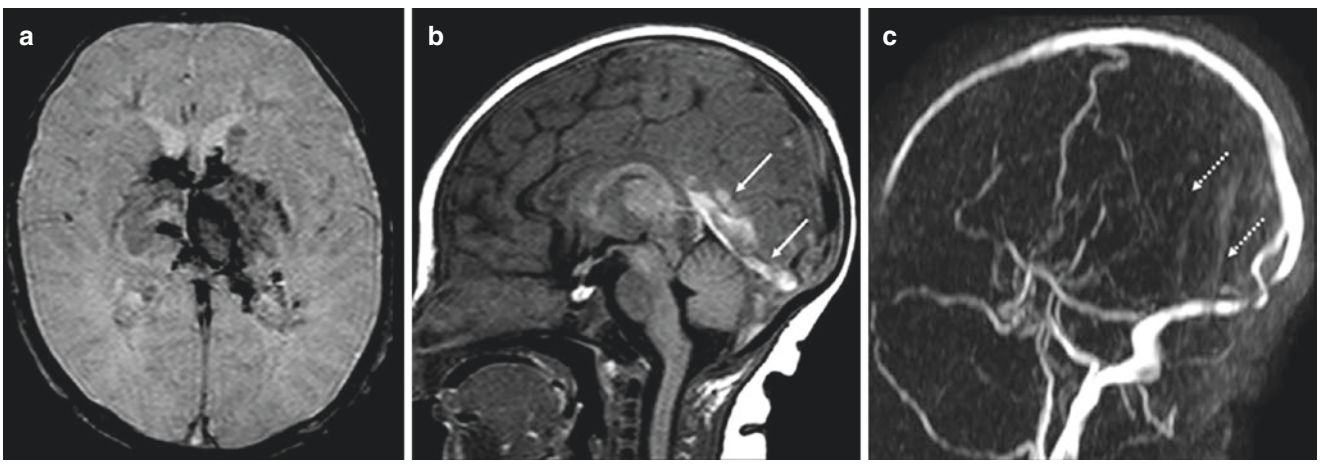

Fig. 14.2 Straight sinus thrombosis in a 6-year-old boy. (**a**) Gradient-echo T2*-weighted image shows venous infarction involving both thalami with a focal hemorrhage on the left side. (**b**) Sagittal T1-weighted image shows subacute thrombus in straight sinus (arrows). (**c**) MR venogram confirms absent flow in straight sinus (dashed arrows)

14.3 Intracranial Hemorrhages

Excluding head trauma, an important cause of intracranial hemorrhages (ICH), spontaneous ICH is a rare, but potentially fatal event in childhood [11]. The most common causes of ICH include vascular malformations (e.g., AVM, cavernous angiomas, and aneurysms), hematological diseases (e.g., coagulopathies and thrombocytopenias), brain tumors, and rare entities (e.g., Moyamoya disease). In addition, ICH may be observed as a complication of AIS and VS. Higher morbidity and mortality are seen particularly in infratentorial hemorrhages, aneurysms, children younger than 3 years, and children with underlying hematological disorders [11]. The presenting symptoms are non-specific, especially in children younger than 3 years of age, including headache, vomiting, impaired consciousness, convulsions, and focal neurological defects. Subacute courses are frequent and result in delayed diagnosis.

US, CT, and MRI may be used depending on the child's age, clinical presentation, location, and availability. ICH follows a well-defined evolution, and neuroimaging appearance varies with the stage of hematoma: hyperacute (<12 h), acute (12 h to 2 days), early subacute (2–7 days), late subacute (8 days to a month), and chronic (more than 1 month) [12] (Table 14.1).

Table 14.1 Evolution of intra-parenchymal hematoma characteristics on CT and MRI

Hematoma age/ phase	CT	T1-weighted MRI	T2-weighted MRI
Hyperacute (<12 h)	Isodense	Iso/ hypointense	Hyperintense
Acute (12 h–2d)	Increasing density	Iso/ hypointense	Hypointense
Early subacute (2–7 days)	Increased density	Hypointense	Hypointense
Late subacute (8d–1m)	Decreasing density	Hyperintense	Hyperintense
Chronic (>1m)	Hypodense	Hypointense	Hyperintense (hypointense rim)

h hours, *d* days, *m* months

On US, exact differentiation between each phase of the hematoma is limited. Acute hematomas typically appear as iso- or hyperechogenic focal mass lesions. Progressively they become centrally hypoechogenic with decreased mass effect, and in the chronic stage, hematomas may dissolve, leaving a hypoechoic CSF-filled brain defect.

On CT, early hyperacute hematomas are isodense compared to normal brain tissue, and consequently it can be difficult to identify hyperacute hematomas. However, intravenous contrast injection may increase the sensitivity of CT in detecting these lesions. Progressive blood clot retraction increases the density of hematomas in the early subacute phases (Fig. 14.3a), while late subacute hematomas show decreasing density due to progressive red blood cell lysis. In the chronic phase, progressive hematoma resorption results in a hypodense, CSF-filled brain lesion.

Similar temporal signal changes are observed on MRI (Fig. 14.3b–d) depending on (1) magnetic susceptibility effects of the evolving blood products and the different oxidation states of the iron within the hemoglobin, (2) magnetic field strength, and (3) applied MRI sequence. T2*GRE and SWI should be included due to the paramagnetic susceptibility effects of hemosiderin. DWI also shows a well-defined temporal evolution of hematomas [12]. Contrast-enhanced sequences and MRA and MRV sequences may be used to exclude vascular malformations or neoplasms as underlying causes of ICH.

Key Points

- ICH is a frequent cause of high morbidity and mortality in children.
- Depending on the etiology and evolutionary phase of the ICH, CT, and MRI imaging characteristics change over time.

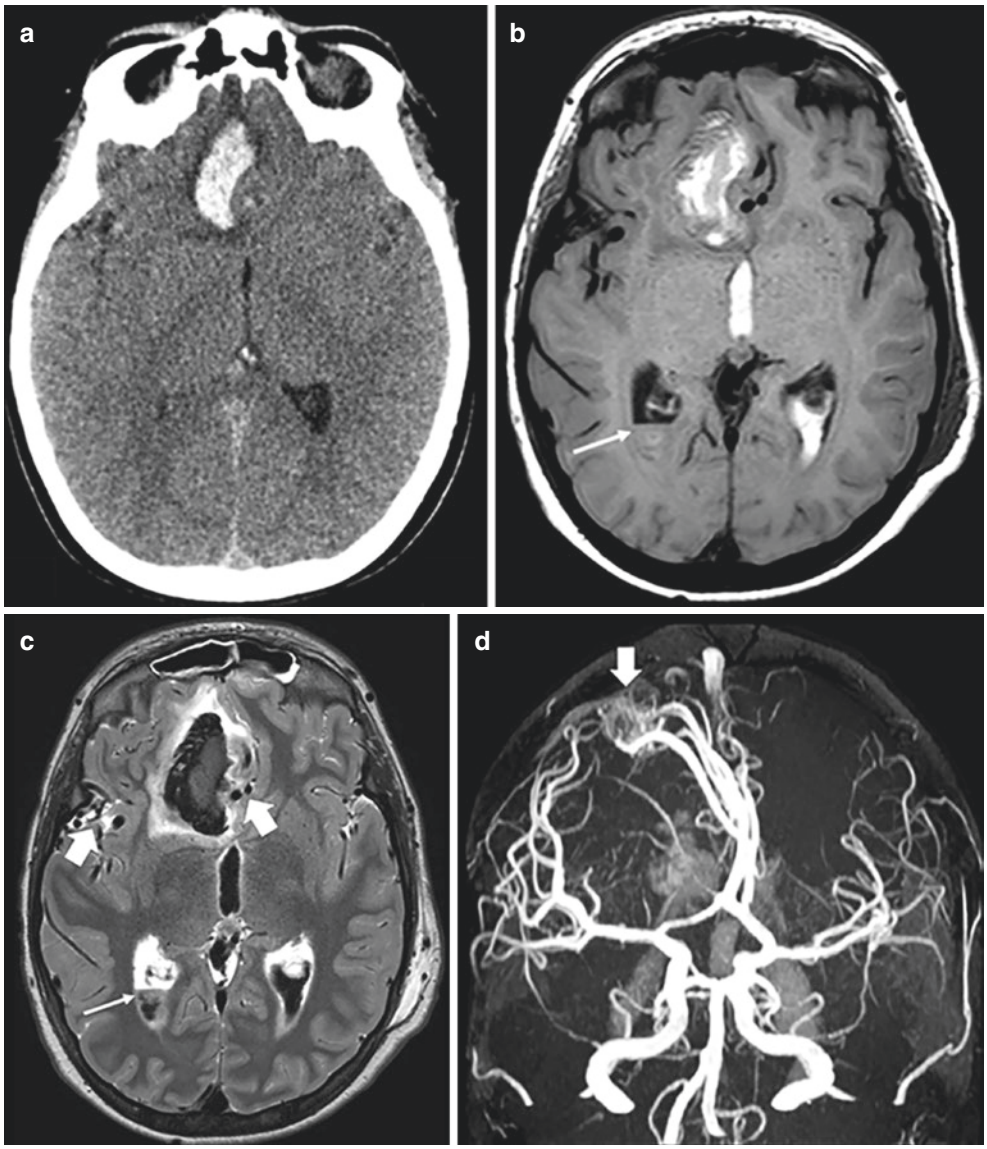

Fig. 14.3 Ruptured AVM in a 13-year-old boy. (**a**) Axial CT scan shows hyperdense hematoma in right frontal lobe. (**b**) Axial T1-weighted image and (**c**) Axial T2-weighted image shows hematoma is mainly T1-hyperintense and T2-hypointense, consistent with an early subacute phase (intracellular methemoglobin) and elicits vasogenic edema. Notice intraventricular penetration, with dependent layering in the right occipital horn (thin arrow, **b** and **c**) suggesting more recent bleeding (isointense in T1 and T2—oxyhemoglobin). Also notice prominent vascularity consisting of hypertrophied anterior and middle cerebral artery branches (thick arrows, **c**). (**d**) 3D TOF MR angiogram confirms hypertrophied arterial afferent converging on to a nidus (arrow)

14.4 Seizures

Acute seizures are the most common neurologic emergencies in neonates [13]. Seizures are sudden, uncontrolled bursts of electrical activity in the brain that cause changes in behavior, movements, and levels of consciousness. The increased susceptibility of the neonatal brain is related to age-dependent physiologic features of the developing brain that lead to increased neuronal excitation and decreased inhibition. Seizures are most commonly caused by acute brain injuries. Hypoxic/ischemic encephalopathy is responsible for seizures in up to 90% of full-term newborns [2], followed by intracranial hemorrhages, stroke, infections, acute hydrocephalus, electrolyte and metabolic disturbances, leukodystrophies, and congenital CNS malformations [13]. In a minority of children, seizures may be unprovoked and secondary to epilepsy.

Timely and accurate diagnosis is critical for optimizing management and determining prognosis.

Initial assessment includes general physical and neurological examination, laboratory tests, and EEG monitoring. Neuroimaging is required depending on testing results, and MRI without contrast (Fig. 14.4) is the preferred imaging modality because it has a superior anatomic resolution and is more detailed than CT in assessing potential causative entities. Functional MRI provides further information and helps localize the epileptogenic foci even in cases where structural MRI is normal [14]. In neonates, US may be performed as an initial imaging investigation. Neuroimaging is not indicated in children with febrile seizures but should be considered for

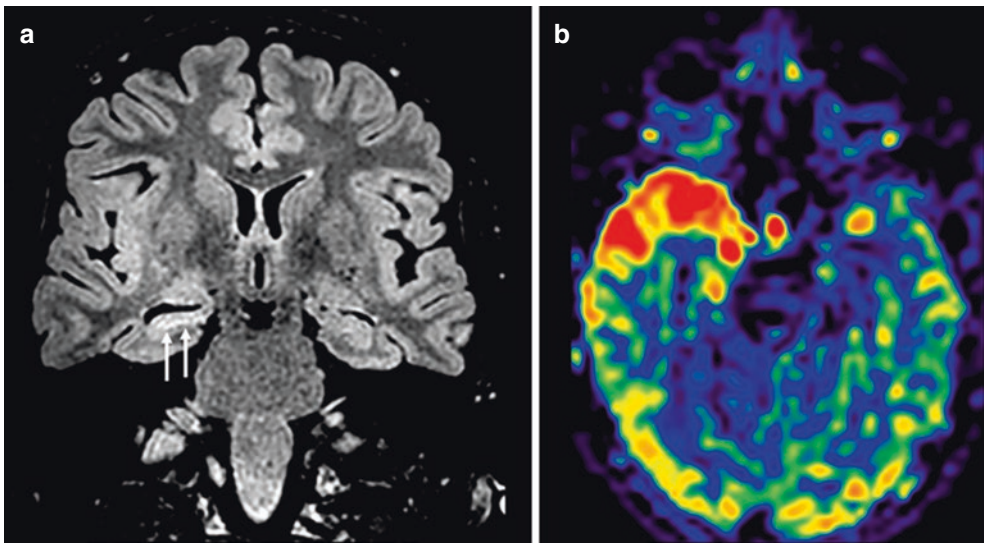

Fig. 14.4 Peri-ictal changes in a 15-year-old girl with ongoing complex partial seizures. (**a**) Coronal FLAIR image shows right hippocampal sclerosis. (**b**) Colorized arterial spin labeling (ASL) image shows markedly increased perfusion signal in the anterior temporal lobe, which normalized at 7-day follow-up (not shown)

complex seizures or new-onset seizures without a febrile illness [3]. The aim is to detect focal disorders that need immediate intervention.

Key Points

- Seizures are the most common neurologic emergencies of children presenting to the ER and EEG is mandatory.
- Neuroimaging should be saved for cases with history of long-time seizures with focal onset and focal neurological deficits, and in neonates.

14.5 Trauma

Traumatic brain injury (TBI) is a leading cause of disability and mortality in children. Birth trauma resulting from instrumental delivery is responsible for almost all cases of neonatal traumas. In contrast, non-accidental trauma is the most common cause of traumatic death in the first 2 years of life. In toddlers and adolescents, falls and motor vehicle accidents represent the main cause of TBJ [15]. Clinical assessment is often challenging, and the emergency work-up requires a multidisciplinary approach. Pediatric TBJ may occur because of the initial trauma (primary injury) or as a complication of it (secondary injury). Secondary injuries are largely preventable and treatable and imaging plays a key role in limiting their extension [15].

Primary injuries can be extra-cranial (epidural, subdural hemorrhage, subarachnoid, and intraventricular hemorrhage), intra-axial (cortical contusion, intracerebral hematoma, and diffuse axonal injury), or vascular (carotid cavernous fistula, arteriovenous dural fistula, vascular dissection, and pseudoaneurysm) (Fig. 14.5). Secondary injuries may be acute (diffuse cerebral edema, brain herniation, and infection) or chronic (hydrocephalus, encephalomalacia, cerebrospinal fluid leak, and leptomeningeal cyst). Multiple imaging modalities are currently available depending on the type, quality, degree, and time of injury. The minimal standard MRI protocol includes (1) sagittal 3D T1WI, which allows multiplanar reconstruction, (2) axial T2WI, (3) axial DWI or DTI sequence, and (4) axial SWI sequence from the skull base to the vertex [16].

In an emergency setting, the presence of hemorrhages of different ages, retinal hemorrhages, change or inconsistency in the reported trauma history, delay in seeking medical care by the caregivers, overall poor care, or when the encountered imaging findings are inconsistent with the trauma history, non-accidental head trauma should be suspected. The most common mechanisms of injury in abusive head trauma include blunt impact, acceleration/deceleration injuries, neck strangulation, and chest compression that may lead to reduced blood flow and consequent brain ischemia (Fig. 14.6). In the setting of accidental and non-accidental trauma, elevated lactate, reduced N-acetyl aspartate, and elevated choline/related compounds on MRS have been demonstrated to indicate poor outcomes by some authors [17].

Further diagnostic workup should include spinal cord injuries, but this topic will be covered in a separate paragraph dedicated to spinal emergencies.

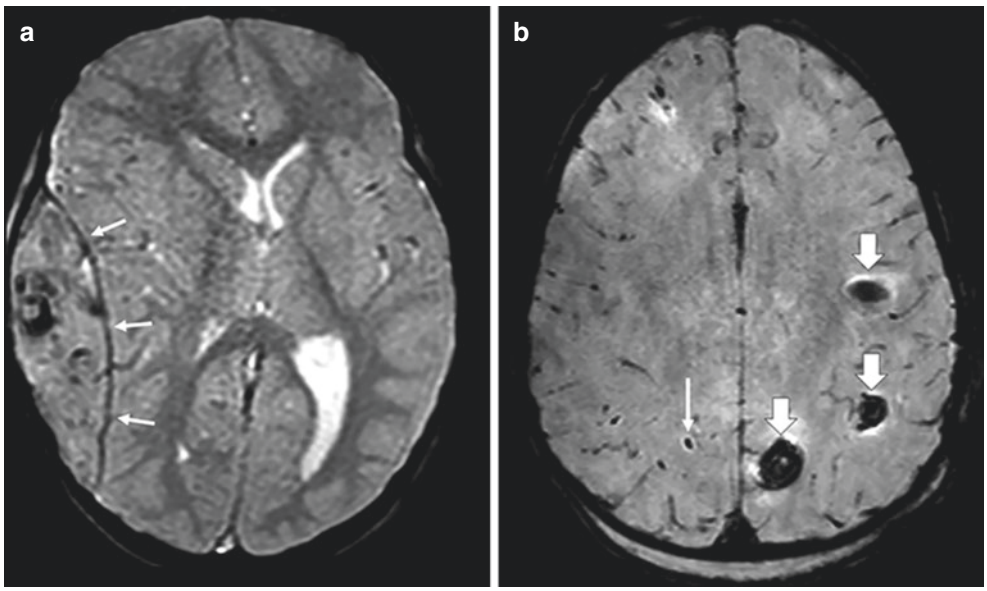

Fig. 14.5 Accidental head trauma. (**a**) Axial gradient-echo T2*-weighted image in a 3-year-old girl shows epidural hematoma; notice typical biconvex shape of the blood collection (arrows). (**b**) Axial susceptibility-weighted image in a 3-year-old boy who was in a motor vehicle accident shows large hemorrhagic (thick arrows) and punctate (thin arrow) areas of traumatic axonal damage

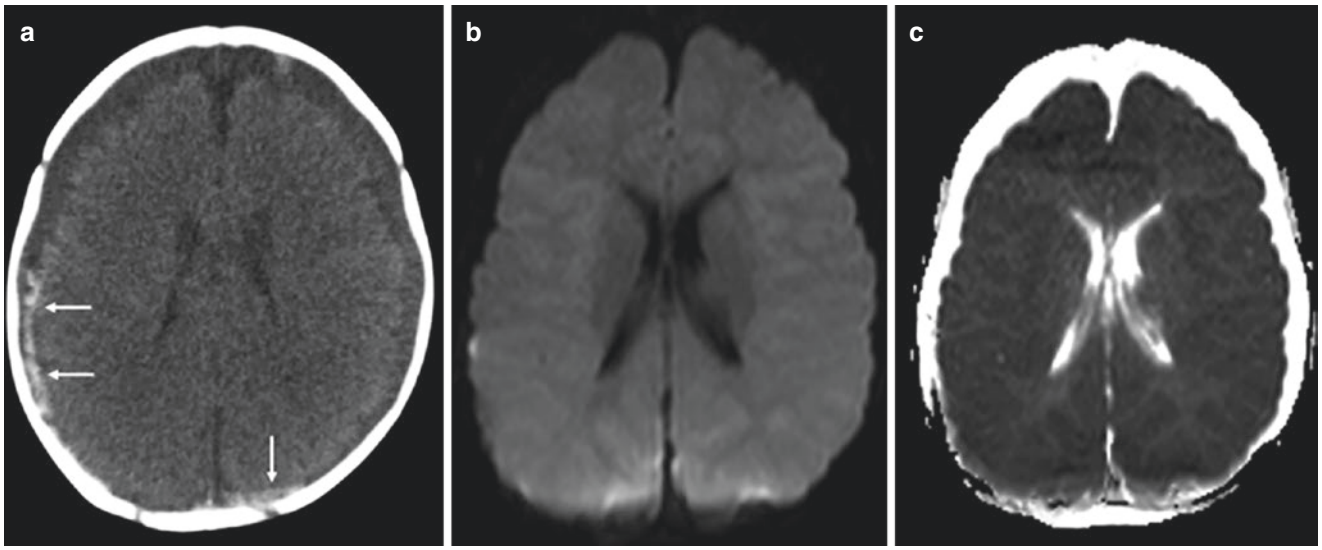

Fig. 14.6 Abusive head trauma in a 2-month-old boy who was found unresponsive. (**a**) Axial CT scan shows diffuse cerebral edema with loss of demarcation between gray and white matter; mixed density subdural hematoma with hyperdense components (arrows) are seen bilaterally. (**b**) Diffusion-weighted image and (**c**) corresponding ADC map confirm diffuse cytotoxic edema with profoundly restricted diffusion

Key Points

- Trauma is a leading cause of disability and mortality in children and depending on the etiology of TBJ and age of the child, CT and MRI are the primary modality of choice.
- It is important to keep in mind child abuse when dealing with trauma in children in an emergency setting. Particularly, in presence of delay in medical care, overall poor care, or when the encountered imaging findings are inconsistent with the trauma history.

14.6 Infections

Infections of the central nervous system (CNS) can be life-threatening if not promptly diagnosed and treated. When a child presents to the ER with non-specific signs and symptoms, including headache, fever, altered mental status, and behavioral changes, CNS infections should be suspected and considered in the differential diagnoses [18]. In an emergency setting, in conjunction with medical history and clinical and laboratory findings, neuroimaging accurately determines brain involvement and suggests the diagnosis in cases where this has yet to be established [19].

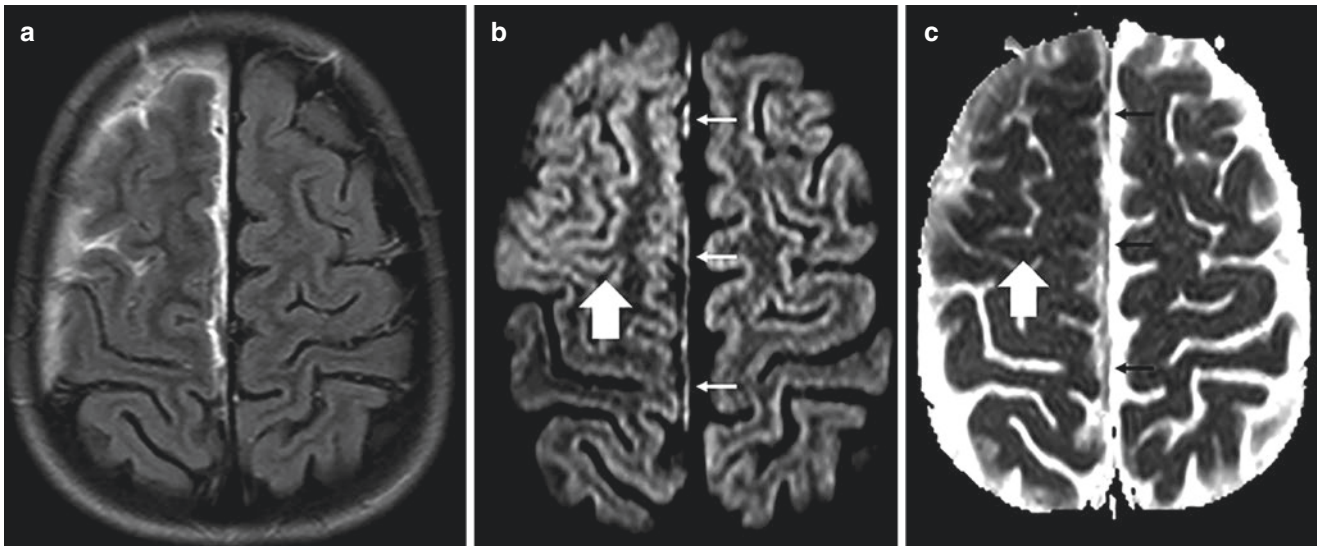

Fig. 14.7 Bacterial meningoencephalitis in a 7-year-old girl. (**a**) Axial post-contrast FLAIR image shows enhancement of the right frontal subarachnoid spaces, consistent with arachnoiditis. (**b**) Axial diffusion-weighted image and (**c**) corresponding ADC map reveal restricted diffusion affecting the right frontal lobe (thick arrow), suggesting cerebritis. Also notice restricted diffusion of the dura along the falx (thin arrows)

Meningitis is the most common form of CNS infection, and bacterial meningitis is usually associated with a higher morbidity and mortality than viral meningitis [18, 19]. Group B streptococcus and *Neisseria meningitidis* are the most common causative bacteria in the neonatal period and in children older than 2 years, respectively [19]. In most cases, meningitis occurs by hematogenous spread from bacterial infection outside the CNS. However, it may also develop from an adjacent infective focus (e.g., sinusitis, mastoiditis), or direct invasion after a skull fracture.

The diagnosis of meningitis is clinical and based on laboratory analysis. A lumbar puncture demonstrating increased white blood cells in the CSF confirms the diagnosis. Neuroimaging is generally used to evaluate complications, including hydrocephalus, subdural effusion, empyema, cerebritis, brain abscess, and infarctions (including strokes secondary to vasculitis). MRI is the modality of choice and T1- and T2WI, fluid-attenuated inversion recovery (FLAIR), SWI, and DWI are especially helpful in evaluating the full extension of CNS affection [2] (Fig. 14.7).

Cerebritis and brain abscesses typically present with focal neurological deficits, seizures, visual defects, personality changes, and more generic symptoms (e.g., fever, headache, nausea/vomiting). Classic MR imaging findings of an abscess include a contrast-enhanced rim surrounding a necrotic core. They appear hyperintense on T1WI, hypointense on T2 WI, and show true restricted diffusion on DWI (Fig. 14.8).

Pyogenic ventriculitis is a life-threatening form of infection. Contrast-enhanced MRIs typically show ependymal enhancement (Fig. 14.9) and restricted diffusion on DWI with reduced ADC values, especially if pus is collecting within the ventricles [20].

Subdural empyema is an infected fluid collection between the dura and arachnoidea. It is a neurologic emergency that can progress rapidly, increasing intracranial pressure and leading to coma and death within 24–48 h. They appear hyperintense on T2WI, isointense on T1WI and show peripheral enhancement [3] (Fig. 14.10).

Acute viral encephalitis is the most common type of encephalitis and often occurs with meningeal involvement [21]. It is caused by direct cytotoxic neurotropic action of viruses that reach the CNS by hematogenous spread or retrograde axonal transport.

Herpes simplex virus (HSV) type 1 is the most common causative agent [3]. Infection predominately affects the limbic system, particularly the medial temporal and inferior frontal lobes [22]. MRI is superior to CT in detecting the often-subtle edema involving the limbic system in the initial stages (Fig. 14.11). In later stages, progressing edema and hemorrhagic necrosis are seen in the hippocampus, cingulate gyrus, and insular and frontobasal regions [19]. Gyral swelling and edema appear hypointense on TWI, hyperintense on T2W- and FLAIR images, and show restricted diffusion on DWI. It is important to note that the enhancement on DWI may disappear within 2 weeks, whereas the hyperintense signal on T2W/FLAIR images may last longer [19].

HSV type 2 encephalitis occurs more often in neonates and immunocompromised patients. Imaging findings of HSV-2 encephalitis are much less uniform, and lesions can

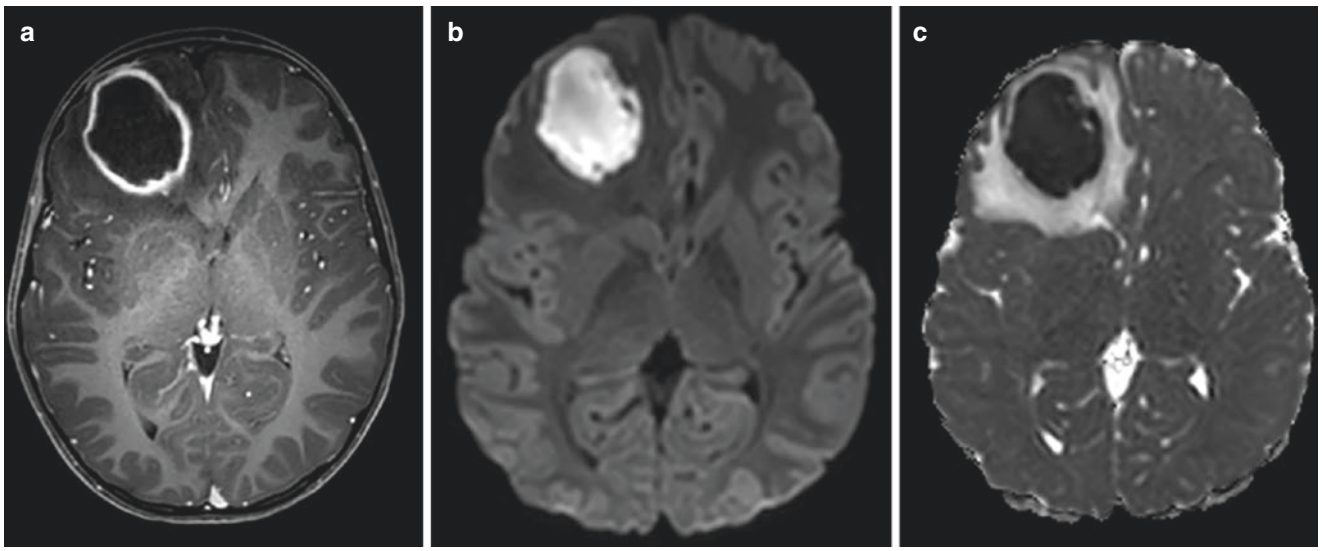

Fig. 14.8 Pyogenic abscess in an 8-year-old boy. (**a**) Axial post-contrast T1-weighted image shows ring-enhancing mass in the right frontal lobe. (**b**) Axial diffusion-weighted image and (**c**) corresponding ADC map show profoundly restricted diffusion consistent with a pyogenic core. Notice marked vasogenic edema surrounding the abscess, characterized by increased diffusion

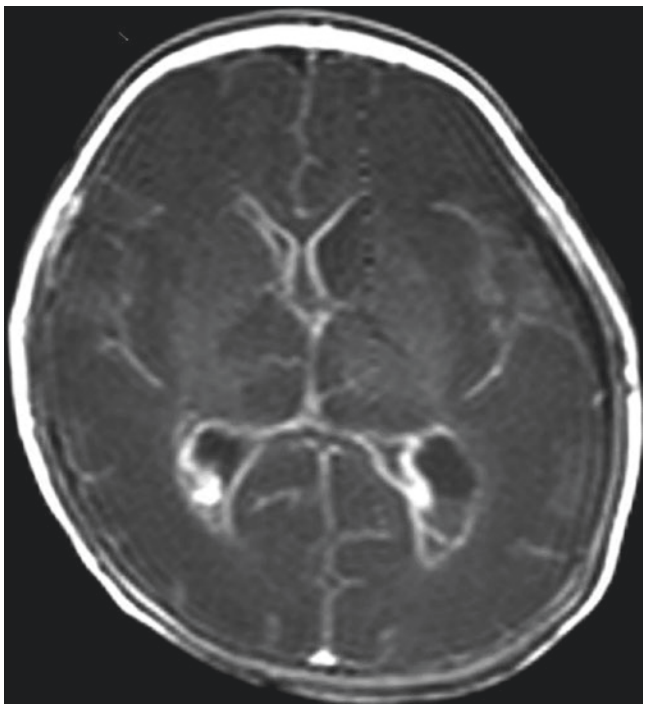

Fig. 14.9 Ventriculitis in a 1-day preterm newborn. Axial post-contrast enhancement shows diffuse ependymal enhancement. Also notice enhancing subarachnoid spaces due to concurrent meningitis

be either multifocal or limited to the temporal lobes, cerebellum, or brainstem [23]. Watershed distribution ischemic injury may be described in areas remote from the primary herpetic lesions (Fig. 14.12). Hemorrhage is less often compared with HSV-1 encephalitis [24].

Key Points

- The diagnosis of uncomplicated meningitis is based on laboratory testing and neuroimaging is limited to evaluate threating complications including empyema or parenchymal infections. MRI is the diagnostic modality of choice.
- Acute viral encephalitis is most commonly caused by HSV type 1 and neuroimaging shows typical lesions involving the limbic system on initial stages.

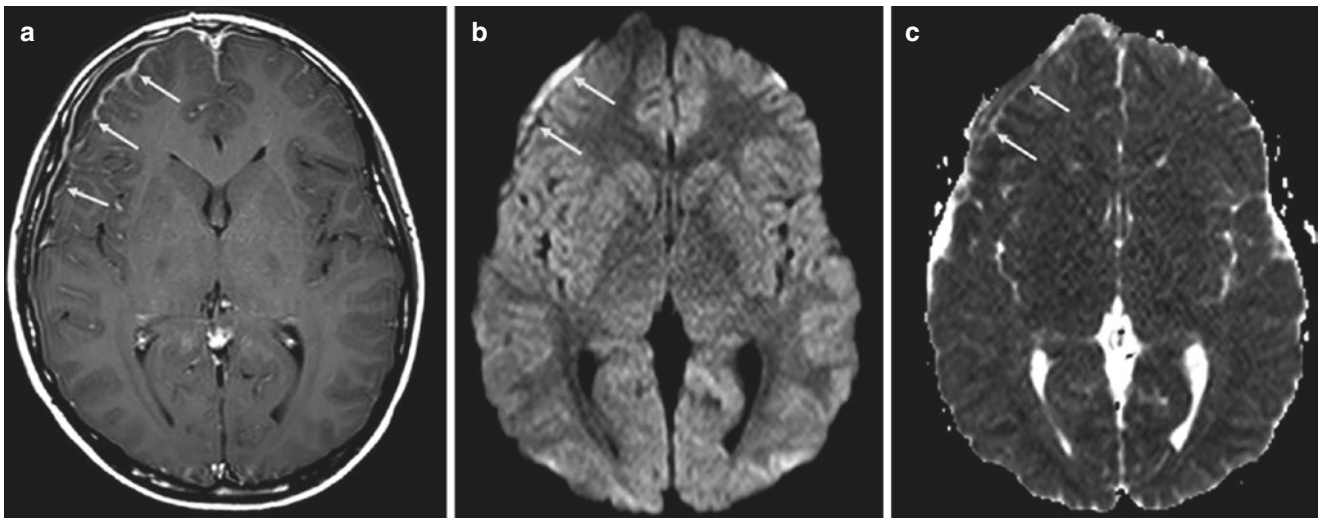

Fig. 14.10 Subdural empyema in a 9-year-old boy presenting with fever and convulsions. (**a**) Axial post-contrast T1-weighted image shows enhancing arachnoid (arrows) underlying a subdural collection along the surface of the right frontal lobe. (**b**) Axial diffusion-weighted image and (**c**) corresponding ADC map shows the subdural collection gives restricted diffusion consistent with a pyogenic collection

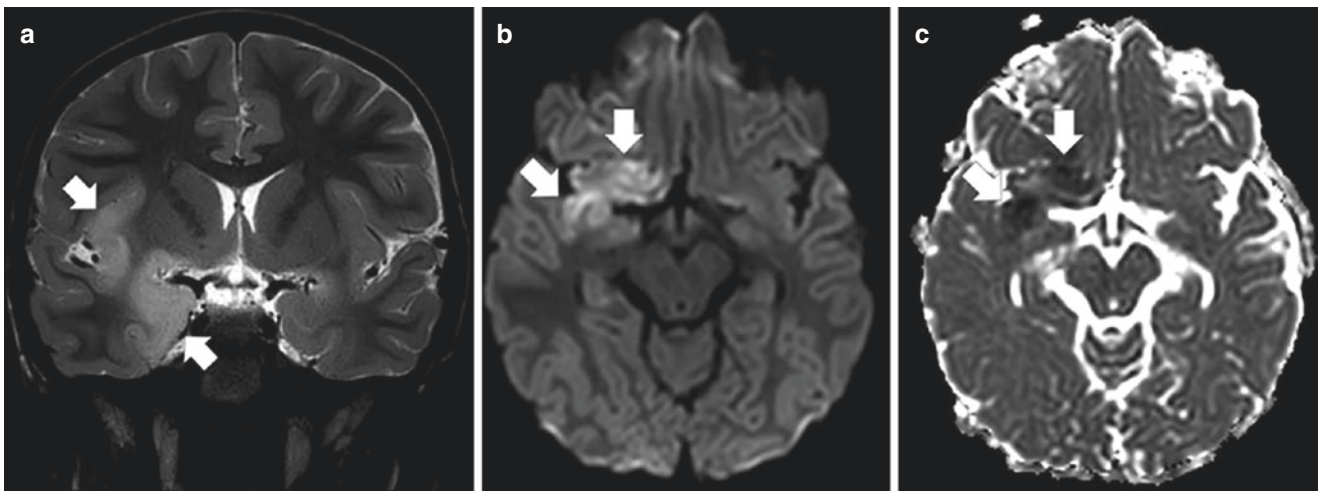

Fig. 14.11 Encephalitis due to herpes simplex virus-1 in a 3-year-old boy presenting with seizures, hemiparesis, and impairment of consciousness. (**a**) Coronal T2-weighted image shows swollen, hyperintense cortex in the right mesial temporal lobe and insula. (**b**) Axial diffusion-weighted image and (**c**) corresponding ADC map show restricted diffusion in the involved cortex

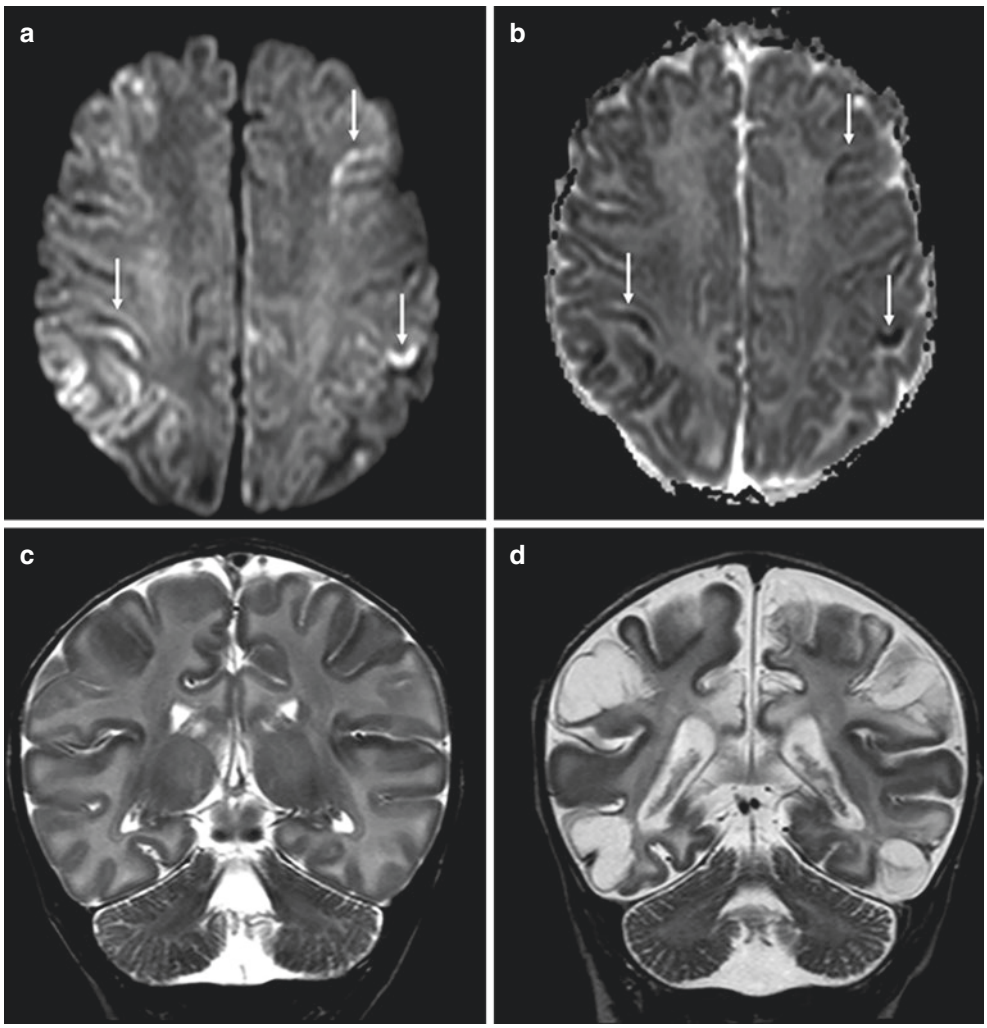

Fig. 14.12 Encephalitis due to herpes simplex virus-2 in a 15-day newborn presenting with seizures. (**a**) Axial diffusion-weighted image and (**b**) corresponding ADC map show multifocal areas of restricted diffusion involving the cortical gray matter of both cerebral hemispheres. (**c**) Coronal T2-weighted image is unrevealing; however, (**d**) coronal T2-weighted image obtained after 1 month reveals severe brain damage with extensive cortico-subcortical liquefaction

14.7 Metabolic and Toxic Imbalances

Neurometabolic imbalances are characterized by acute onset in a previously healthy child, and manifest with progressive and rapid worsening of the symptoms. Laboratory testing is fundamental and neuroimaging may provide further information to diagnose or evaluate the degree of brain injury and estimate the prognosis.

Diabetic ketoacidosis (DKA) is a serious complication in children with type 1 diabetes that may lead to significant long-term neurological morbidity or mortality. Neuroimaging typically shows brain edema and, in severe cases, may cause focal stroke and subfalcine or transtentorial brain herniation. Focal infarctions typically occur in the mesial basal ganglia, thalamus, periaqueductal gray matter, and dorsal pontine nuclei [25] (Fig. 14.13).

Methotrexate (MTX)-induced leukoencephalopathy is an adverse neurologic effect that may occur in children with leukemia after receiving MTX both intravenously and intrathecally [26]. Children may present with stroke-like symptoms, including headache, confusion, lethargy, seizures, transient paresis, aphasia, and dysarthria. MRI shows diffuse T2/FLAIR hyperintensities in the deep white matter of the centrum semiovale and initial sparing of the subcortical U-fibers (Fig. 14.14). DWI will often demonstrate cytotoxic edema as areas of restricted diffusion across multiple vascular territories in the involved regions. These lesions typically resolve once MTX is withdrawn [27].

Osmotic demyelination syndrome (ODS) is an acute demyelination process associated with electrolyte imbalance. Most frequently, it occurs after too rapid medical correction of hyponatremia and is rare in children. It may present with central pontine myelinolysis (CPM) or extrapontine myelinolysis (EPM). On MRI, CPM shows T2-hyperintensity in the central pons, with sparing of the ventrolateral pons, tegmentum, and corticospinal tracts. This produces a characteristic trident-shaped, bat-winged, or piglet appearance. The lesion is hypointense on T1 with no mass

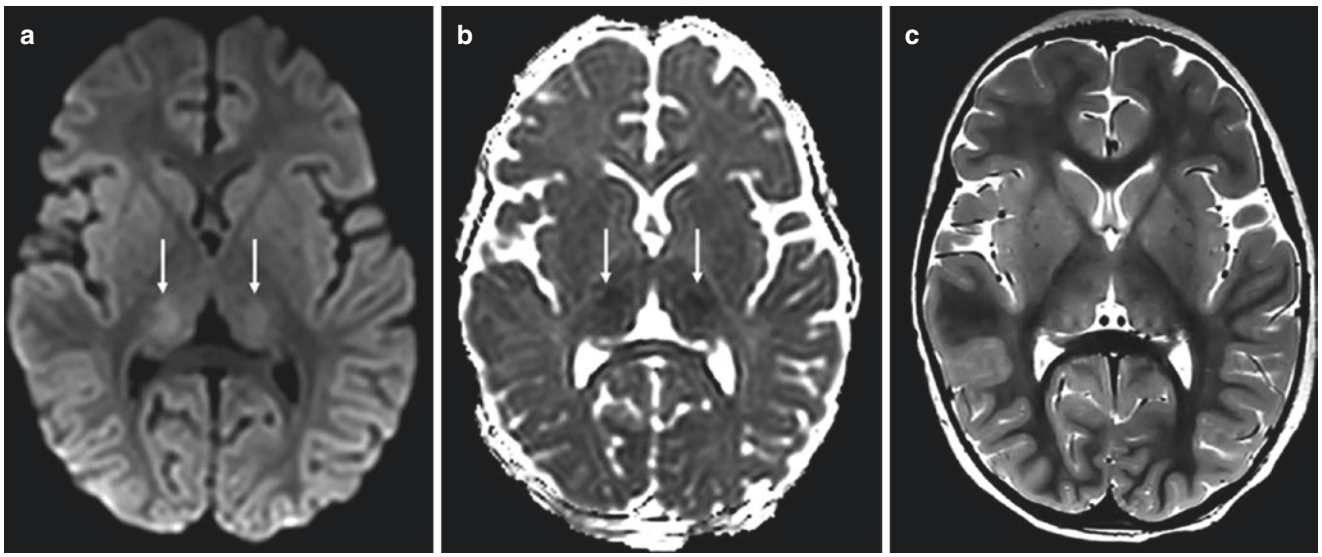

Fig. 14.13 Diabetic ketoacidosis in a 1-year-old boy presenting with impaired consciousness. (**a**) Axial diffusion-weighted image and (**b**) corresponding ADC map reveal reduced diffusivity in both thalami (arrows). (**c**) Axial T2-weighted image shows corresponding mild, ill-defined hyperintensity

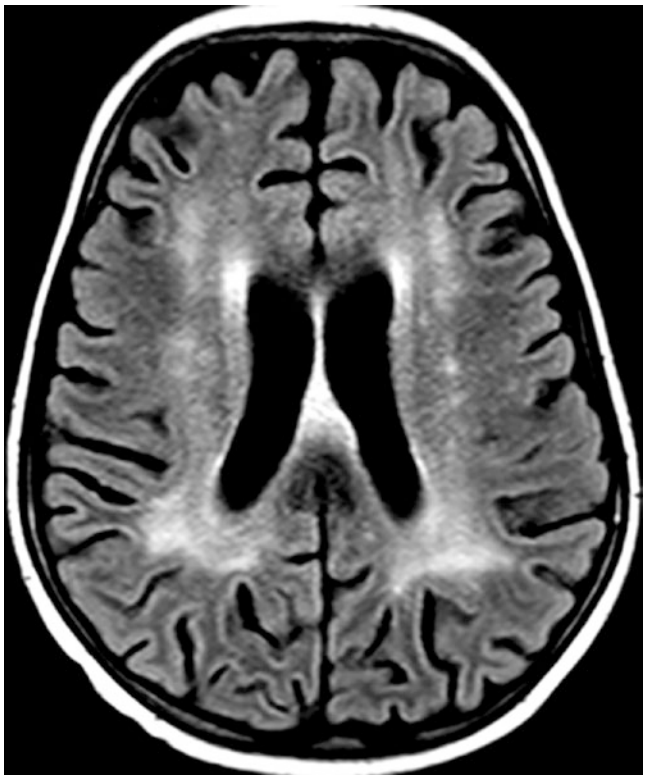

Fig. 14.14 Acute toxic leucoencephalopathy in a 9-year-old girl with relapse of acute myeloid leukemia treated with methotrexate and intrathecal steroids. Axial FLAIR image shows ill-defined, partially confluent, mostly symmetric hyperintense areas involving the deep white matter of the bilateral centrum semiovale

effect. In EPM, lesions are usually bilateral and noted over cerebellar peduncles, globus pallidus, thalamus, lateral geniculate body, putamen, external and extreme capsule, splenium of corpus callosum, and supratentorial white matter [27] (Fig. 14.15).

Key Points

- In children with toxic/metabolic imbalances, along with laboratory testing, neuroimaging may provide further information to make the diagnosis and to evaluate the degree of brain injury and estimate the prognosis.
- Localization of the lesions depends on the etiology and MRI is typically the diagnostic modality of choice.

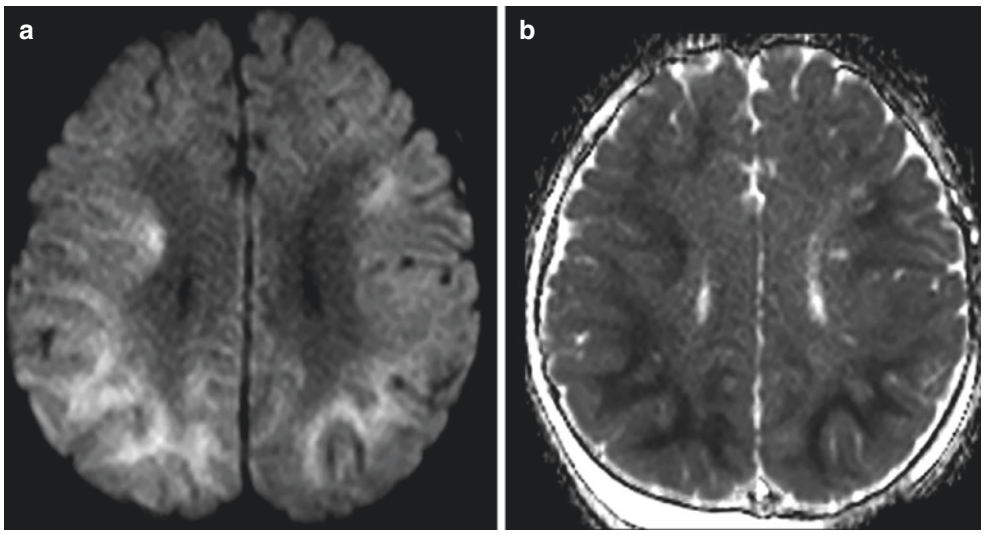

Fig. 14.15 Extrapontine myelinolysis in a 2.5-year-old patient with recent hypothalamic surgery and presenting with diabetes insipidus and hyponatremia. (**a**) Axial diffusion-weighted image and (**b**) corresponding ADC map shows extensive areas of restricted diffusion at level of the subcortical white matter bilaterally. Complete resolution was noted at 6-month follow-up (not shown)

14.8 Autoimmune Pathologies

Acute disseminated encephalomyelitis (ADEM) is an immune-mediated inflammatory disease characterized by perivenular lymphocytic inflammation with acute demyelination in the CNS that occurs 1–3 weeks following an upper respiratory infection or vaccination. It is a monophasic disease with sudden onset, causing rapidly progressive impairment of consciousness and multifocal neurologic symptoms. Brain MRI shows focal or multifocal, patchy, and bilateral T2W and FLAIR hyperintense areas, mainly affecting the basal nuclei and white matter [3, 19] (Fig. 14.16). The cerebellum and spinal cord may also be involved, but these rarely present as isolated lesions without an accompanying lesion in the brain [28]. Acute hemorrhagic encephalomyelitis (AHEM) is a hyperacute variant of ADEM that rapidly progresses to coma and may be fatal. Similarly to ADEM, AHEM occurs in response to a preceding infection or immunization and presents with symmetric multifocal neurologic deficits, headache, and seizures [29].

ADEM is typically a diagnosis of exclusion; therefore, demyelinating disorders, particularly multiple sclerosis (MS), and myelin oligodendrocyte glycoprotein-associated disease (MOGAD) should be considered in the differential diagnosis.

MS most commonly affects adults, whereas ADEM presents at an early age. Furthermore, follow-up MRIs in MS usually demonstrate new and often asymptomatic demyelinating lesions [30].

Neuromyelitis optica spectrum disorder (NMOSD) is an inflammatory disorder of the CNS characterized by severe, immune-mediated demyelination, and axonal damage predominantly targeting optic nerves and the spinal cord. Serum titer of anti-aquaporin-4 antibodies is an important biomarker for the diagnosis. However, MRI also plays an important role in the differential diagnosis of NMOSD, particularly with MS. Longitudinally extending myelitis (>3 vertebral segments) appears hyperintense on T2WI and hypointense on T1W sequences. In most cases, it involves the cervical and upper thoracic spinal cord segments with a central gray matter predominance [31] (Fig. 14.17a, b). Optic nerve sheath thickening, with T2-hyperintensity and gadolinium enhancement on T1-weighted sequences (Fig. 14.17c) has been described in both NMOSD and MS. In contrast to MS, in NMSOD, the optic nerve involvement is typically bilateral and lesions are seen in the optic chiasm as well. Brain lesions include periependymal T2- and FLAIR hyperintense diencephalic lesions surrounding the third ventricles and cerebral aqueduct, dorsal brainstem lesions adjacent to the fourth ventricle, periependymal lesions surrounding the lateral ventricles as well as hemispheric white matter and corticospinal tracts involving lesions [31].

Key Points

- Children with autoimmune CNS diseases may present to the ER with acute signs and symptoms.
- In addition to CSF and serum laboratory findings, brain and spinal cord MRI is helpful to differentiate MS from ADEM and NMSOD.

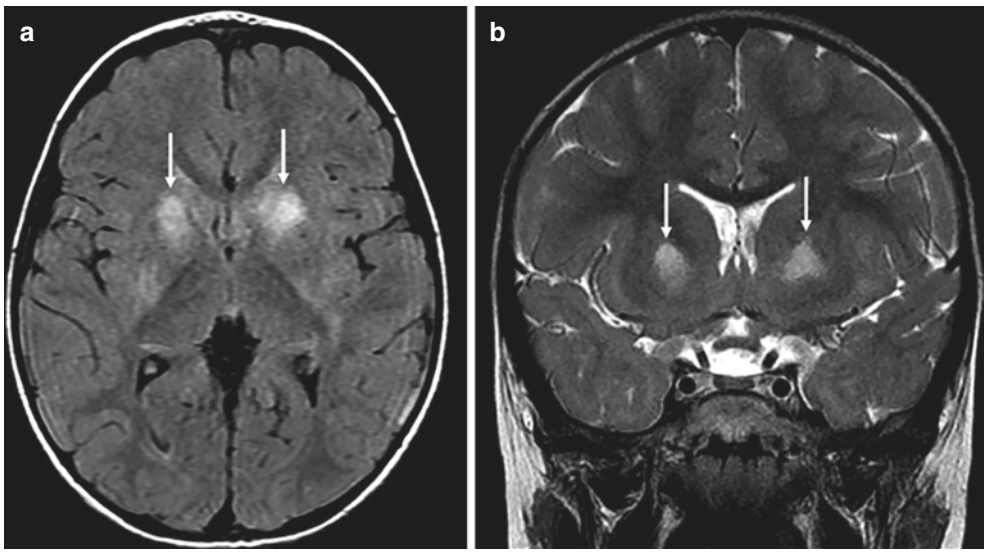

Fig. 14.16 Acute disseminated encephalomyelitis (ADEM) in a 2-year-old girl presenting with reduced consciousness 2 weeks after an upper airway infection. (**a**) Axial FLAIR image and (**b**) coronal T2-weighted image show hyperintense signal involving the striatum bilaterally

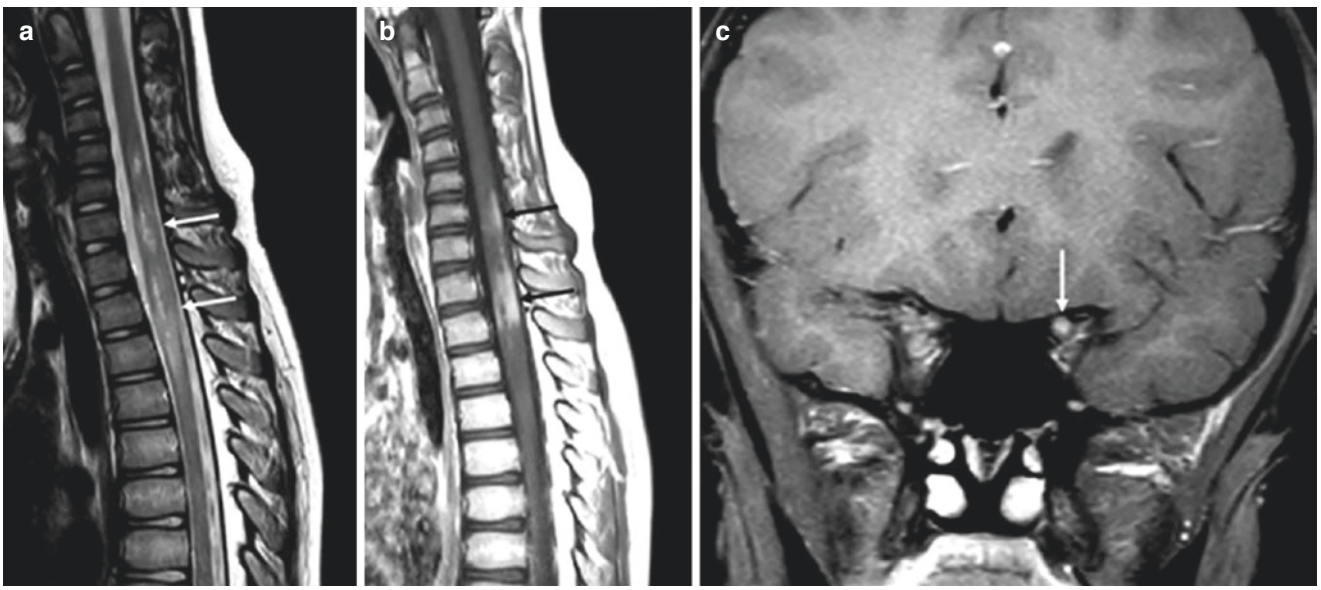

Fig. 14.17 Neuromyelitis optica in a 10-year-old boy presenting with tetraparesis and visual impairment. (**a**) Sagittal T2-weighted image and (**b**) post-contrast sagittal T1-weighted image shows swollen, hyperintense cervico-thoracic spinal cord with irregular enhancement. (**c**) Post-contrast, fat-suppressed coronal T1-weighted image shows enhancing intraforaminal segment of the left optic nerve

14.9 Acute Facial Nerve Palsy

Facial nerve palsy (FNP) is frequently presented in pediatric emergency departments. Bell's palsy is an idiopathic or potentially post-viral peripheral form of FNP and constitutes the most common cause of FNP in the pediatric population [32]. Other causes include (1) infectious diseases (e.g., otitis media, mastoiditis, Herpes Zoster, Lyme disease); (2) neoplasms/malignancies (e.g., posterior fossa tumors, parotid gland tumors, leukemia, lymphoma); (3) trauma/nerve compression (perinatal trauma, temporal bone fractures, otic barotrauma); (4) congenital (facial nerve malformation, cardiofacial syndrome), (5) inflammatory; and (6) metabolic disorders [33]. FNP results from a peripheral lesion of the facial nerve or a central lesion involving the upper motor neuron due to damage above the facial nucleus. Clinical symptoms include immobility of the brow, incomplete lid closure, drooping of the corner of the mouth, impaired closure of the lips, dry eye, hyperacusis, impaired taste, and pain around the ear [34].

Bell's palsy is an exclusion diagnosis, and imaging is not typically required to confirm the diagnosis. Imaging is indi-

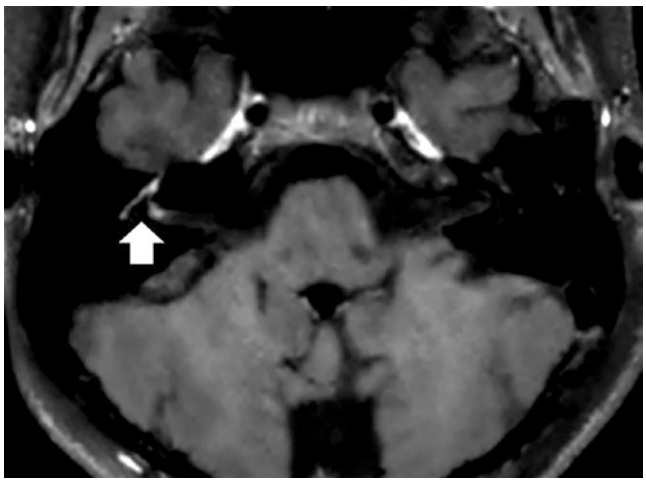

Fig. 14.18 Facial neuritis in a 9-year-old girl presenting with peripheral facial paralysis. Post-contrast axial T1-weighted image shows enhancement of geniculate and tympanic segments of right facial nerve

cated only in the presence of symptoms inconsistent with Bell's palsy, such as progressive onset or paralyses, multiple cranial nerve involvement, no signs of recovery after 3/6 months from the onset, recurrent paralysis, suspected malignancy, or in traumatic cases [33, 35]. Contrast-enhanced CT and MRI are beneficial in these cases, allowing evaluation of all portions of the facial nerve [33, 36] (Fig. 14.18).

Key Points

- FNP is a frequent presentation in pediatric emergency department and, idiopathic FNP (Bell's palsy) constitutes the most common cause of FNP in children.
- Bell's palsy is an exclusion diagnosis and neuroimaging is indicated only in the presence of recurrent paralysis, suspected malignancy, progressive onset, and multiple cranial nerve involvement.

14.10 Hydrocephalus

Hydrocephalus in children can occur due to CSF overproduction, obstruction of the CSF flow (communicating and non-communicating type), or decreased CSF absorption due to dysfunctional Pachionic granulations. Pediatric patients may present to the emergency department with symptoms related to acute hydrocephalus or secondary to the complications of shunted hydrocephalus. Clinical presentation varies with age and includes irritability, vomiting, and bulging of the fontanelle in infants. In older children, headache, vomiting, diplopia, or papilledema can be seen [37]. Neuroimaging is necessary to identify the underlying causes, and MRI is the diagnostic modality of choice. Recommended sequences include axial thin-section T2WI, followed by coronal and sagittal reconstruction [3]. The most common imaging findings (Fig. 14.19) include (1) ventriculomegaly, (2) enlargement of the third ventricular recesses and lateral ventricular and/or temporal horns, (3) decreased mamillopontine distance and frontal horn angle, (4) thinning and elevation of the corpus callosum, (5) normal or narrowed cortical sulci, (6) periventricular white matter interstitial edema, and (7) aqueductal flow void phenomenon on T2W images (a sign of communicating hydrocephalus) [38].

Complications of shunted hydrocephalus include infections and shunt malfunctions. CT is often the primary imaging modality because it is readily available and can assess the length of the shunt catheter, the causes of the obstruction and the site of tubing disconnections or migration [3]. Scout views can be useful for detecting possible shunt disconnections below or outside the scanned region that may be missed on axial or reconstructed cross/sectional CT images [39].

CSF cultures are usually sufficient for the diagnosis in children with infections due to a shunted hydrocephalus. Imaging may be indicated in children that along with fever, altered mental status, nausea, and vomiting, present with abdominal symptoms from a peritoneal CSF pseudo-cyst. Abdominal US or CT are helpful in these cases [2].

Key Points

- Children may present to the ER with acute finding of hydrocephalus or complications of shunted hydrocephalus including shunt malfunction and infection.
- MRI and CT are useful to differentiate the etiology.

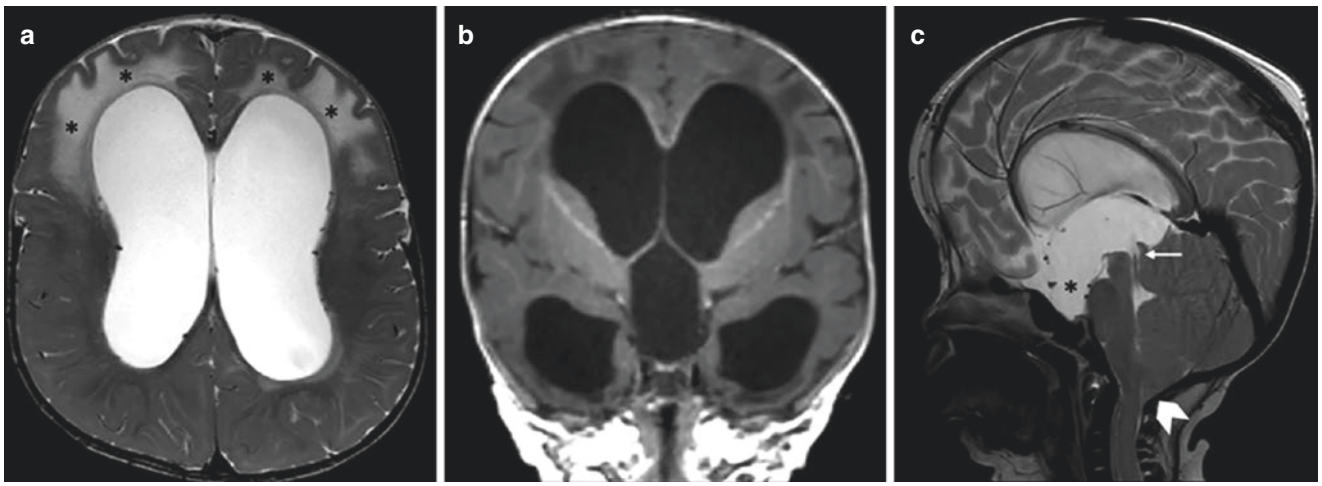

Fig. 14.19 Obstructive hydrocephalus in a 1-year-old girl with Chiari I deformity presenting with severe headache and a large head. (**a**) Axial T2-weighted image shows marked ventriculomegaly with periventricular edema (asterisks) suggesting an unbalanced condition which may herald rapid clinical deterioration. (**b**) Coronal T1-weighted image shows commensurate dilatation of the temporal and frontal horns, an important element in the differentiation from ex-vacuo ventriculomegaly. (**c**) Sagittal T2-weighted image shows marked ectasia of the third ventricle, prominently including the anterior recesses (asterisk); the aqueduct is patent as shown by flow artifact (arrow), while the cerebellar tonsils are protruded into the foramen magnum (arrowhead)

14.11 Drowning/Near Drowning

Anoxic brain injury is a well-known consequence of drowning and may cause severe lifelong neurologic disabilities or, in severe cases, death. Irreversible injury of the hippocampi, basal ganglia, and cerebral cortex have been demonstrated within 4–10 min following the anoxic event. Therefore, early imaging is important for prompt detection of injuries and treatment. CT is useful to detect head and neck trauma, cerebral edema, and loss of white matter differentiation; however, a normal appearing brain parenchyma on early CT does not exclude brain injury. DWI/DTI with ADC mapping on MRI imaging improves the ability to detect brain injuries within minutes of the incident and predict the outcome. Cortical and deep brain DWI abnormalities, and lower ADC values are associated with poor outcomes (Fig. 14.20). MRS best predicts outcome 3–4 days after drowning [40].

Key Points

- Pediatric brain is extremely sensitive to hypoxia and irreversible brain injury develops within 4–10 min after drowning.
- DWI/DTI and ADC mapping on MRI are the most useful sequences to detect brain injuries in children after drowning.

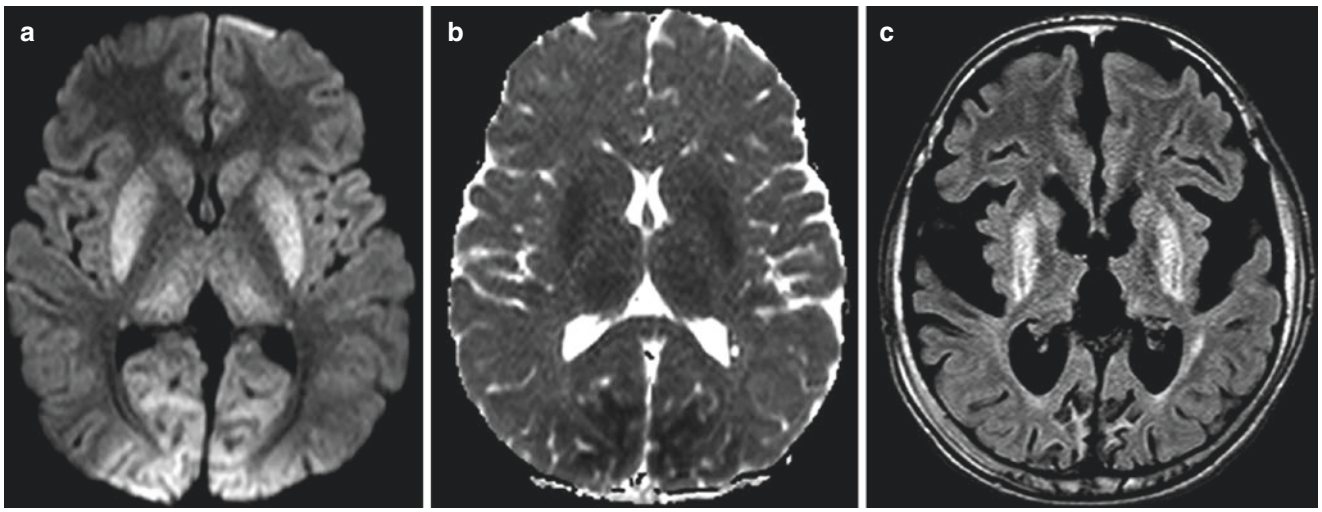

Fig. 14.20 Near drowning in an 8-year-old boy. (**a**) Axial diffusion-weighted image and (**b**) corresponding ADC map show restricted diffusion in the bilateral caudate nucleus, putamen, thalamus, and occipital cortex. (**c**) Axial FLAIR image after 3 months shows profound cerebral atrophy and residual signal changes in the putamen, optic radiations, and calcarine cortex bilaterally

14.12 Spinal Cord Emergencies

Spinal cord emergencies manifest with acute onset of upper and/or lower extremity symptoms (paresis/paralysis), sphincter dysfunction, difficulty walking, and paresthesias in the extremities. Trauma is the most common etiology for pediatric spinal emergencies; other causes include infectious and inflammatory diseases and, more rarely, neoplasms and vascular emergencies [41].

Pediatric spinal trauma is often secondary to motor vehicle accidents, sports-related injuries, falls, and child abuse [42]. Injury location depends on the age of the patients (more often cervical spine due to anatomic differences in the developing spine) [41] and on the injury mechanism (L2–L4 levels in seat-belt flexion-distraction injury) (Fig. 14.21). CT is typically the first imaging study to be performed, and multiplanar reconstructions of the spine show fractures and dislocations with high sensitivity. MRI is necessary to detect spinal cord injuries, epidural hematomas, and intramedullary hemorrhages and allows a better depiction of the adjacent soft tissue [16].

Apart from ADEM, NMOSD, or MS that may involve the spine (previously described), acute spine inflammatory diseases include Guillain-Barre syndromé. It is a demyelinating polyradiculopathy, typically preceded by Campylobacter and Cytomegalovirus infection. MRI demonstrates nonnodular enhancement of the spinal nerve roots, particularly of the cauda equine (Fig. 14.22). Similar lesions and T2-hyperintensities in the anterior spinal cord gray matter are found in acute infection from West Nile virus, too. It results in extremity weakness due to viral tropism for the anterior horn cells of the spinal cord [41].

Key Points

- Trauma is the leading cause of spinal acute emergencies in pediatric population followed by infectious and inflammatory diseases.
- CT is typically the first imaging study to be performed showing fractures and dislocations with high sensitivity. MRI is necessary to detect spinal cord injuries, epidural hematomas, intramedullary hemorrhages, and allows a better depiction of the adjacent soft tissue.

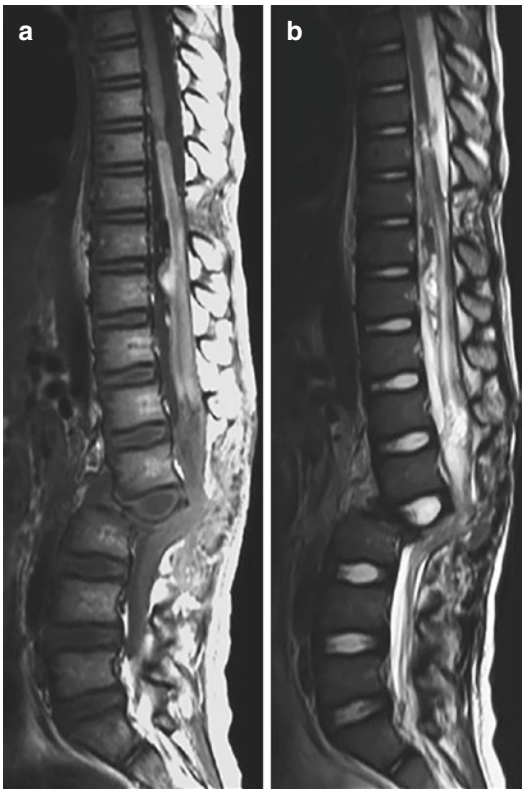

Fig. 14.21 Chance fracture due to seat-belt injury in a 7-year-old boy. (**a**) Sagittal T1-weighted image and (**b**) sagittal T2-weighted image show posteriorly dislocated L2 vertebral body and underlying L2–3 disc with resulting kyphosis and extensive intraspinal hemorrhage, involving both the spinal cord and thecal sac

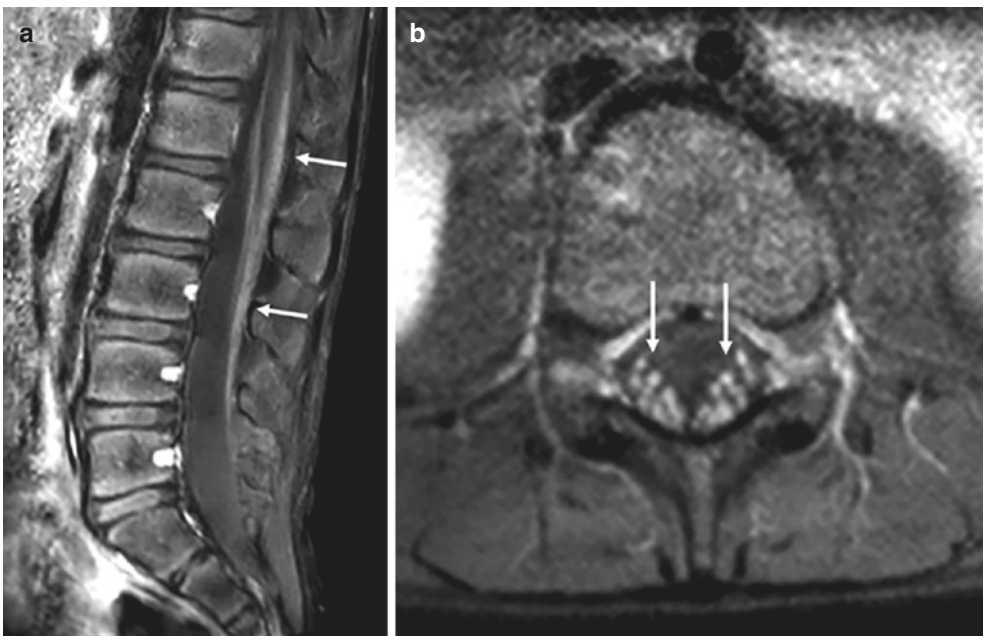

Fig. 14.22 Guillain-Barré syndrome in an 8-year-old boy presenting with lower limb hypotonia and hyporeflexia. (**a**) Sagittal and (**b**) axial post-contrast fat-suppressed T1-weighted images show enhancing cauda equina nerve roots

14.13 Conclusion

Children with acute neurological emergencies present to the ER with a wide spectrum of symptoms and signs. Several challenges are faced by radiologists. Firstly, they should decide if neuroimaging is required emergently. Typically, neuroimaging is necessary in cases where it is difficult to gather good and reliable patient history or adequate physical/neurological examination. Secondly, radiologists must be able to choose the most appropriate imaging modality. CT and MRI are used most often in the emergency setting. Lastly, radiologists should be familiar with the age-specific anatomy and maturational changes related to the pediatric age in order to provide reliable differential diagnosis and prompt patient management.

Take-Home Messages

- Children with acute neurological emergencies may present to the ER with non-specific symptoms or subtle findings.
- Radiology is an integral part of the ER and the choice of the appropriate imaging modality depends on the symptoms, age of the patient, and institution availability.
- Radiologists should be familiar with the CNS biologic and morphologic changes related to the pediatric age in order to provide reliable differential diagnosis and prompt patient management.

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