Infectious and Inflammatory Diseases of the Spine in Children

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Abstract

Inflammatory and infectious disorders of the spine and spinal cord are less common in children than in adults. They are classified according to the involved spinal compartments into those predominantly affecting (i) the spinal cord, (ii) the nerve roots and meninges, and (iii) the vertebrae, discs, and epidural space. This chapter will highlight the major entities in this heterogeneous group of disorders, with an emphasis on the use of magnetic resonance imaging (MRI) and, when needed, computed tomography (CT) and radiographs in their diagnosis.

Introduction

Inflammatory and infectious disorders of the spine and spinal cord are less common in children than in adults. The most common infectious spinal disorder in children is bacterial meningitis, which generally does not require imaging studies and is diagnosed and treated on a clinical and physical examination basis. In bacterial meningitis, imaging studies are reserved for patients suspected of harboring complications from the disease.

For purposes of clarity, inflammatory and infectious spinal disorders in childhood will be divided, according to the affected spine compartment, as follows:

(i) Those predominantly affecting the spinal cord
(ii) Those predominantly affecting the nerve roots and meninges
(iii) Those predominantly affecting the vertebrae, discs, and epidural space

The use of magnetic resonance imaging (MRI) and, when needed, computed tomography (CT) and radiographs will be emphasized. Although many of the disorders discussed in this chapter also show brain abnormalities, discussions will be limited to the abnormalities found in the spine and spinal cord.

Disorders Affecting the Spinal Cord

Inflammatory and Demyelinating Disorders

Acute Transverse Myelitis
Acute transverse myelitis (ATM) is a focal inflammatory disorder of the spinal cord characterized by an acute onset of motor, sensory, and autonomic dysfunction. Individuals of all ages may be affected; approximately 20% of cases of ATM occur in the pediatric age group (Scott et al. 2011), with two peaks of pediatric incidence, between 0 and 2 years of age and 5–17 years of age (Pidcock et al. 2007). Although the terms “acute transverse myelopathy” and “acute transverse myelitis” have often been used

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interchangeably, the former is a broad header that includes idiopathic forms (corresponding to acute transverse myelitis) and forms with known cause, such as postinfectious and postvaccination (i.e., acute disseminated encephalomyelitis, ADEM), ischemic, paraneoplastic, autoimmune, and postirradiation myelitis.

A prodrome of mild illness is common in the few weeks preceding the clinical onset of disease (Wolf et al. 2012). Clinical presentation is with pain (60% of cases), paresthesias, leg weakness, and sphincter dysfunction, all of which progress to nadir between 4 h and 21 days (usually 24 h) following the onset of symptoms. It is believed that stroke-like evolution (i.e., nadir reached earlier than 4 h) indicates vascular causes (spinal cord infarction). Signs and/or symptoms are usually bilateral, though not necessarily symmetric, and there usually is a clearly defined sensory level, as suggested by the term “transverse” indicating the band-like sensory disturbance across the midline that is typically described in this condition (Wolf et al. 2012). Cerebrospinal fluid (CSF) analysis reveals signs of spinal cord inflammation, such as pleocytosis or elevated IgG index. Prognosis of ATM is variable, with 1/3 of patients recovering with few or no sequelae, 1/3 left with moderate degree of permanent disability, and 1/3 having severe disabilities (Transverse Myelitis Consortium Working Group 2002). Rapid progression of signs and symptoms at presentation usually portends a poorer prognosis.

Histopathology studies show intraparenchymal and perivascular lympho-monocytic cellular infiltrates associated with demyelination and neuronal injury. Pathophysiological theories involve molecular mimicry, superantigen effect, humoral-based dysregulation, interleukin 6-mediated toxicity (IL-6), and allergy phenomena with elevated immunoglobulin E (IgE) levels. The end result of these phenomena is ischemia with resulting cord lesions.

The diagnosis of ATM (Table 1) is one of exclusion. The first step is to rule out other conditions including extrinsic spinal compression, ischemia, tumor, arteriovenous malformation, and toxicities (i.e., vitamin B12 deficiency, previous spinal radiation) (Wolf et al. 2012). Differentiation from spinal cord ischemia may be challenging. However, while ischemia typically has a stroke-like presentation, progression to nadir of the neurological deficits is slower in ATM, requiring at least 4 h (Wolf et al. 2012). Moreover, spinal cord ischemia is frequently associated with predisposing conditions, including cardiovascular surgery or fibrocartilaginous embolism. Next, identified causes of ATM such as connective tissue disease, ADEM, multiple sclerosis, and neuromyelitis optica (NMO) must be ascertained. Cases lacking a specific diagnosis are labeled as idiopathic ATM. The role of MRI in this diagnostic approach is clearly paramount. Because the presentation of compressive lesions such as extramedullary tumor or hemorrhage can be identical to ATM (Wolf et al. 2012), an emergent MRI study is warranted. It is wise to perform a complete neuraxial (i.e., spine and brain) MRI study in order to assess possible additional lesions (such as those typical of ADEM or multiple sclerosis) without delay, while minimizing the need for additional sedation.

**Imaging Findings** The spinal MRI study should include the whole spinal axis, including the craniovertebral junction and the bottom of the thecal sac, and should include the intravenous administration of gadolinium chelate. High-resolution sagittal T1- and T2-weighted images should be obtained; sagittal short-tau inversion recovery (STIR) is also extremely useful to detect subtle signal intensity abnormalities of the spinal cord. Optimal slice thickness for sagittal studies should be 3 mm or less. Axial T2-weighted images across lesional areas are very important to determine the cross-sectional extent of the spinal cord involvement, an important element in the differential diagnosis of ATM (Thurnher et al. 2007). Postcontrast images should be acquired in the three planes of space. Fat suppression may increase the conspicuity of contrast enhancement.

MRI criteria for myelitis (Fig. 1) (Tartaglino et al. 1996; Choi et al. 1996) include normal or slightly expanded spinal cord showing diffuse or patchy hyperintensity on T2-weighted images, usually involving
more than three to four vertebral levels in length and more than 2/3 of the cross-sectional area of the spinal cord, with a prevalingly central location which involves both the central gray matter and surrounding white matter. Lesion enhancement after gadolinium administration can be patchy or diffuse and depends on the degree of inflammation and timing of imaging with respect to clinical onset, being more common in the subacute stage of the disease (Thurnher et al. 2007; Grayev et al. 2014). Absence of enhancement does not rule out ATM; in two pediatric studies, absent enhancement was reported in 26 % (Pidcock et al. 2007) and 77 % (Thomas et al. 2012) of cases, respectively. The thoracic cord and conus medullaris are involved more frequently, but cases of a quasi-holocord extension are sometimes encountered (longitudinally extensive transverse myelitis, LETM) (Fig. 2). Diffusion-weighted imaging (DWI) reveals increased diffusivity corresponding to the areas of increased signal on the T2-weighted images (Fig. 1) (Thurnher et al. 2007), while diffusion tensor imaging (DTI) reveals reduced fractional anisotropy (FA) values in the involved spinal cord regions and increased FA values in the periphery of the lesion (Renoux et al. 2006). Brain MRI is normal in cases of idiopathic ATM. The presence of brain lesions, or clinical evidence of encephalopathy, should address the hypothesis of ADEM (see below).

Differentiation from spinal cord ischemia may be challenging on imaging. Typical anterior spinal artery distributions will result in the “snake’s eyes” appearance on axial T2-weighted images due to involvement of the anterior horns of the gray matter. DWI will show areas of restricted diffusion in the case of spinal cord ischemia (Fig. 3).

**Acute Disseminated Encephalomyelitis**

Acute disseminated encephalomyelitis (ADEM) involves a first episode of inflammatory demyelination with polyfocal neurological signs implicating involvement of multiple sites of the CNS. Patients present with a rapid onset of encephalopathy and motor and/or sensory deficits with brainstem signs and

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**Table 1** Criteria for idiopathic ATM

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<th>Inclusion criteria</th>
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<td>Development of sensory, motor, or autonomic dysfunction attributable to the spinal cord</td>
<td>History of previous radiation to the spine within the last 10 years</td>
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<td>Bilateral signs and/or symptoms (though not necessarily symmetric)</td>
<td>Clear arterial distribution or clinical deficit consistent with thrombosis of the anterior spinal artery</td>
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<td>Clearly defined sensory level</td>
<td>Abnormal flow voids on the surface of the spinal cord c/w AVM</td>
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<td>Exclusion of extra-axial compressive etiology by neuroimaging (MRI or myelography; CT of spine not adequate)</td>
<td>Serologic or clinical evidence of connective tissue disease (sarcoidosis, Behçet’s disease, Sjögren’s syndrome, SLE, mixed connective tissue disorder, etc.)a</td>
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<td>Inflammation within the spinal cord demonstrated by CSF pleocytosis or elevated IgG index or gadolinium enhancement. If none of the inflammatory criteria are met at symptom onset, repeat MRI and lumbar puncture evaluation between 2 and 7 days following symptom onset to meet criteria</td>
<td>CNS manifestations of syphilis, Lyme disease, HIV, HTLV-1, <em>Mycoplasma</em>, other viral infections (e.g., HSV-1, HSV-2, VZV, EBV, CMV, HHV-6, enteroviruses)a</td>
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<td>Progression to nadir between 4 h and 21 days following the onset of symptoms (if patient awakens with symptoms, symptoms must become more pronounced from the point of awakening)</td>
<td>Brain MRI abnormalities suggestive of MSa</td>
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| History of clinically apparent optic neuritis |


aDo not exclude disease-associated ATM
Fig. 1  Acute transverse myelitis in a 12-year-old boy. (a) Sagittal T1-weighted image shows a swollen conus medullaris. (b) Sagittal T2-weighted image shows hyperintense signal involving the swollen conus medullaris and extending cranially to the thoracic cord. (c) Gd-enhanced sagittal T1-weighted image shows the affected cord is essentially nonenhancing. (d) Gd-enhanced fat-suppressed sagittal T1-weighted image obtained at a slightly later (10 min) time shows faintly increased enhancement on the posterior aspect of the conus (arrowheads). The pial surfaces of the cord also show enhancement owing to venous congestion. (e) Coronal T2-weighted image confirms significant swelling of the conus medullaris with an ill-defined hyperintense signal that extends cranially in the cord. (f) Axial T2-weighted image obtained at the level of the conus medullaris shows the abnormal signal involves the vast majority of the cross-sectional area of the cord. (g) Gd-enhanced T1-axial weighted image confirms the lesion is essentially nonenhancing. (h) Axial diffusion-weighted image (b 1,000 m/s²) across the conus shows high signal consistent with a T2-shine-through phenomenon. (i) Corresponding ADC map confirms the absence of restriction at this level, consistent with vasogenic edema.
symptoms and ataxia. The presence of encephalopathy (defined as altered behavior or consciousness) is a required criteria for ADEM, and, as such, it is considered as a separate entity from ATM (Yiu et al. 2009).

Most patients with ADEM present in the aftermath of infection. Postinfectious ADEM has been related to viruses such as measles, rubella, chickenpox, mumps, influenza, Epstein–Barr virus, Coxsackie B, cytomegalovirus, herpes simplex virus, hepatitis A virus, and adenoviruses, as well as with *Borrelia, Mycoplasma pneumoniae*, and nonspecific infection of the upper respiratory tract. Postvaccination ADEM can be induced by several vaccines, including polio, rabies, smallpox, influenza, rubella, and plasma-derived form of hepatitis B (Smith and Ouvrier 1999). In most patients, postvaccination myelitis is a presumptive diagnosis based on the temporal relationship between vaccine administration and onset of symptoms.

Unlike idiopathic ATM, ADEM typically is characterized by extensive involvement of the brain. About 40% of patients with ADEM will also show lesions in their spinal cord (Tartaglino et al. 1996), but the actual proportion may be higher since not every patient with brain lesions consistent with ADEM undergoes spinal cord imaging. The process usually is monophasic, i.e., it occurs once in the life of the patient. However, relapsing–remitting forms (i.e., multifocal disseminated encephalomyelitis, MDEM) have also been documented (Gallucci et al. 2001). The disorder commonly begins 1–2 weeks after a viral, and seemingly minor, illness. Often, the initial illness may be subclinical. Once the symptoms are present, they become more obvious 1–2 days after the diagnosis but may progress for up to 2 weeks. CSF analysis may show increased proteins and leukocytosis. The thoracic spinal cord is involved more often than the cervical region. Histologically, there is necrosis and inflammation; perivascular lymphocytic infiltration and demyelination also are present.

**Fig. 2** Longitudinally extensive transverse myelitis in a 5-year-old girl. (a) Sagittal T1-weighted image is unrevealing. (b) Sagittal T2-weighted image shows ill-defined central hyperintensity of the spinal cord involving most of the thoracic cord and conus medullaris (arrowheads). There is no significant swelling of the cord in this case. (c) Axial T2*-weighted image confirms hyperintensity of the central portion of the spinal cord (arrowheads), exceeding 2/3 of the cross-sectional area of the cord.
Imaging Findings MRI is the best method to assess these patients. T2-weighted images may show multiple, more or less well-defined or demarcated areas of increased signal intensity within the cord (Fig. 4) (Tartaglino et al. 1995a; Rossi 2008). Some of these regions may be confined to the gray matter, others are located in the white matter, and some involve both (Fig. 5). Holocord involvement is possible. Segmental disease generally involves two to three vertebral bodies in length and may expand the cord slightly. In ADEM, generally there is no enhancement after gadolinium administration, while enhancement is not uncommon in idiopathic ATM (Tartaglino et al. 1996). In the latter condition, enhancement of the cauda equina may also be seen, suggesting that transverse myelitis and Guillain–Barré syndrome may have a similar etiology. It has been suggested that the following MRI findings may be helpful in distinguishing idiopathic ATM from ADEM: normal size or segmental enlargement of the cord, most commonly thoracic; central hyperintensity in T2-weighted images affecting three to four vertebral levels; central dot into the core of hyperintensity; and focal nodular or diffuse contrast enhancement at the periphery of the spinal cord (Choi et al. 1996). However, the single most common differential feature is the presence of brain involvement, as evidenced by clinical signs of encephalopathy and MRI evidence of signal abnormalities, in ADEM but not in ATM.

Multiple Sclerosis Multiple sclerosis (MS) is a chronic inflammatory demyelinating disease of the CNS characterized by immune-mediated inflammation and progressive neurodegeneration and burdened by significant neurological impairment. Early onset (i.e., below age 18 years) MS accounts for 2–10 % of all MS cases
Spinal cord involvement was reported to occur in about 50% of patients who underwent a comprehensive brain and spinal cord MRI study at the time of presentation (Hummel et al. 2013). The current revision of the McDonald criteria includes the spinal cord among the 2 of 4 areas of the CNS that are required to fulfill dissemination in space (DIS) (Polman et al. 2011); therefore, spinal cord imaging is paramount in all patients suspected of harboring MS. In one study (Hummel et al. 2013), the McDonald criteria for DIS were satisfied in 9% of cases only after the spinal MRI was taken into account. Spinal cord plaques can be single or multiple, occur preferentially in the dorsolateral cord, and are prevailingly found in the cervicothoracic cord, although predilection for the cervical segment has been reported in the early stages of the disease (Thurnher et al. 2007).

**Imaging Findings** The MRI findings in childhood and juvenile MS mimic those of adult-onset MS (Tartaglino et al. 1995b). T2-weighted images show one or more elongated, poorly marginated, hyperintense intramedullary lesions (Fig. 6). STIR sequence may increase the conspicuity of the lesions and is therefore strongly advised. Typical lesions are relatively small, involving only a portion of the transverse diameter of the spinal cord and not spanning longer than two vertebral levels in craniocaudal length (Hummel et al. 2013; Verhey et al. 2013); this marks an important difference from lesions of ATM that are more typically longer in length and larger in terms of cross-sectional involvement of the spinal cord (Thomas et al. 2012). Acute demyelinating lesions may display mass effect and enhance after gadolinium administration (Fig. 6). Tumefactive plaques in the spinal cord have been reported in association with swelling and MR signal changes mimicking a neoplasm (Glasier et al. 1995).
Neuromyelitis Optica

Neuromyelitis optica (NMO), formerly known as Devic’s disease, is a rare, severe, monophasic or multiphasic demyelinating disease of the CNS that preferentially affects the optic nerves and spinal cord. The revised diagnostic criteria for NMO (Krupp et al. 2007) require the presence of optic neuritis.

Fig. 5 Acute disseminated encephalomyelitis with predominant spinal involvement in a 3-year-old girl with negative NMO serology. (a) Sagittal T2-weighted image shows longitudinally extensive involvement of the cervicothoracic cord, which appears swollen and hyperintense. (b) Gd-enhanced sagittal T1-weighted image shows absence of enhancement. (c) Axial T2-weighted image shows extensive cross-sectional abnormality of the spinal cord with involvement of both gray matter horns and white matter columns. (d) Axial FLAIR image of the brain shows abnormal hyperintensity at the left medial thalamus and massa intermedia (arrow). The remainder of the brain was normal, as well as the optic nerves. (e) Sagittal T2-weighted image obtained at a 3-month follow-up after treatment with steroids and intravenous immunoglobulins shows normalized findings. Brain imaging was also normal at this time (not shown). There was no recurrence over a 5-year follow-up.

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and acute myelitis, associated with either a spinal MRI lesion extending over three or more segments or positive NMO serology. It has been shown that the presence of at least two of three laboratory findings, (i) contiguous spinal cord MRI lesion extending over >3 vertebral segments, (ii) brain MRI not meeting criteria for MS, and (iii) NMO-IgG seropositive status, is 99 % sensitive and 90 % specific for NMO (Wingerchuk et al. 2007).

The relationship between NMO and MS has been the source of enduring debate in the literature, and NMO has often been misdiagnosed as a severe form of MS in the past. Recently, the identification of a specific serologic pattern (Lennon et al. 2005) has helped to define NMO as a spectrum of disease beyond the classic definition of acute transverse myelitis and optic neuritis and to establish NMO as a separate disease from MS and other demyelinating disorders (Wingerchuk et al. 2007). The neuromyelitis optica immunoglobulin G (NMO-IgG) is an autoantibody found in the serum of patients affected by NMO, with a 91–100 % specificity and 75–90 % sensitivity. NMO-IgG binds to aquaporin-4 (AQP4), which is the main channel that regulates water homeostasis in the CNS, mainly located around cerebral microvessels, pia mater, and Virchow–Robin sheaths. Immunopathological studies of active NMO demyelinating lesions have revealed loss of AQP4 and vasculocentric distribution of immune complexes corresponding

**Fig. 6** Multiple sclerosis in a 13-year-old girl. (a) Sagittal STIR image shows several hyperintense areas involving the spinal cord at multiple levels (arrowheads). Notice that lesions are focal and that the individual lesion involves less than three (in this peculiar case, less than one) vertebral levels in length, marking an important difference from both ATM and ADEM. (b) Gd-enhanced sagittal T1-weighted image shows enhancement only involves some of those areas, satisfying the criteria of dissemination in time. (c) Axial FLAIR image of the brain shows several areas of abnormal hyperintensity, some with a tumefactive appearance. The presence of lesions in subcortical, juxtaventricular, and spinal cord areas satisfies the criteria of dissemination in space. (d) Gd-enhanced axial T1-weighted image again shows enhancement of several lesions (arrows); however, one area adjacent to the right frontal horn (arrowhead) does not enhance, again corroborating dissemination in time.
to the normal expression of AQP4 in the endfeet of astrocytes (Misu et al. 2006; Roemer et al. 2007). Disruption of water homeostasis results in demyelination and necrosis with little or no inflammation.

Most affected patients are females, with a 9:1 gender predilection over males (Wingerchuk et al. 2007). While mostly a disease of adults, NMO occasionally is seen in children. Typically, optic neuritis is severe, painful, more commonly unilateral than bilateral, and not simultaneous with ATM. Attacks of optic neuritis precede ATM in 80% of cases (usually by less than 3 months), whereas ATM precedes optic neuritis in 20% of cases. Clinical findings in patients with myelitis prominently include severe symmetric paraplegia, sensory loss below the lesion, and bladder dysfunction.

Atypical forms of NMO have recently been identified in which NMO-IgG seropositivity is found in patients with isolated longitudinally extensive transverse myelitis (LETM), monophasic or recurrent isolated optic neuritis, and brainstem encephalitis. Furthermore, extensive brain involvement with tumefactive lesions reminiscent of tumorlike plaques of MS has been described (Lotze et al. 2008). The terms “NMO spectrum disorder” and “autoimmune AQP4 channelopathy” have been coined to describe this host of conditions that encompass a larger group of patients than the classical NMO criteria (Trebst et al. 2014). Interestingly, it appears that the majority of children with an eventual diagnosis of NMO will present with isolated LETM, optic neuritis, or brainstem encephalitis, thereby not meeting the diagnostic criteria at the time of their first presentation and despite NMO-IgG seropositivity.

**Imaging Findings** Imaging of the entire craniospinal axis should always be performed in patients suspected of harboring NMO, regardless of the primary presenting clinical signs and symptoms (Trebst et al. 2014), and the optic nerves and chiasm should be specifically scrutinized by means of high-resolution sequences. Spinal MRI typically shows a picture of LETM, characterized by large, confluent areas of high T2 signal intensity involving the spinal cord extensively (DeLara et al. 1995) (Fig. 7). These lesions are typically longer than three vertebral bodies in a craniocaudal direction; they occasionally involve the entire span of the cord and may enhance with gadolinium administration. Enhancement can be patchy and inhomogeneous and may persist for weeks to months after the onset of the disease (Trebst et al. 2014). NMO is a necrotizing disease, and as such the cord may be expanded and may show cavitation (Fig. 8). Although the zones of necrosis may contain hemorrhage, this is not always evident on imaging studies. Differential diagnosis with intramedullary tumors may be difficult in the presence of extensive necrotic-cystic changes and contrast enhancement (Tortori-Donati et al. 1993) (Fig. 7). Brain imaging of suspected NMO patients will show optic nerve and chiasm involvement (Fig. 8), characterized by swelling, T2 hyperintensity, and contrast enhancement in the acute demyelination phase. While the brain may be unaffected, hypothalamic, periaqueductal gray, and area postrema lesions can be found in the context of NMO, in up to 60% of cases in some studies (Trebst et al. 2014). Several factors, such as the timing of image acquisition, patient age, ethnicity, and aquaporin-4 antibody status, significantly influence the findings at MRI in individual patients (Tackley et al. 2014).

**Spinal Cord Infections**

**Spinal Cord Abscess/Granuloma**

Bacterial spinal cord abscesses are extremely rare (Murphy et al. 1998). Children may account for up to 20–50% of cases in some series. Among causative organisms (Table 2) (Murphy et al. 1998; Friess and Wasenko 1997), *Schistosoma* is particularly common in children. Tuberculosis is also gaining new ground in Western countries. Fungal infections may also involve the spinal cord and produce granulomas. Fungal disease includes *Candida*, *Aspergillus*, and *Nocardia*. These are mostly seen in adults who are immunosuppressed, i.e., organ-transplantation patients or those with acquired immunodeficiency syndrome (AIDS).
Predisposing conditions include congenital heart disease, disorders of the immune system, patients harboring long-term intravascular access lines, underlying spinal cord tumors, and dermal sinuses. Dermal sinuses may give rise to intraspinal abscesses outside and inside of the spinal cord (Dev et al. 1997) (Fig. 9). Most patients have a history of infection elsewhere and spinal cord involvement may be secondary to either hematogenous or lymphatic spread. The process begins as a myelitis and, if left untreated, may progress to frank abscess formation.

**Imaging Findings** MRI shows increased T2 signal intensity and expansion of the cord. A thin hypointense stripe surrounding the lesion indicates a capsule. After contrast administration there is ill-defined marginal enhancement according to the stage of the inflammatory process (Murphy et al. 1998) (Figs. 9 and 10). Fungal disease tends to produce multiple lesions that may be small, solid, or enhance in a ringlike pattern. They are accompanied by high T2 signal intensity extending beyond the area of enhancement. After initiation of treatment, the signal on T2-weighted images decreases and ring enhancement becomes prominent. With adequate therapy, the enhancement slowly resolves.

**Viral Myelitis**

Viral myelitis may be due to poliovirus, herpes zoster infection, and cytomegalovirus (CMV), especially associated with the acquired immunodeficiency syndrome (AIDS).

**Poliomyelitis** Poliomyelitis is very uncommon today (Malzberg et al. 1993). Most cases of polio and polio-like clusters are related to prior vaccination (particularly the “live” type). The risk of developing poliomyelitis after oral vaccination is 1:2,500,000 doses. Poliomyelitis may also be encountered in...
immunosuppressed patients. The spread of the disease is generally via the fecal–oral route. Only seven to ten cases per year are reported in the United States. Most cases of poliovirus infection are asymptomatic, and many present with only nonspecific aseptic meningitis. Only the paralytic type of the disease will be addressed here. This type of the disease predominantly affects the motor neurons. Spinal cord involvement predominates in about 50% of polio patients and is the most common variant of the disease encountered in children.

The classic clinical presentation is that of fever, nuchal rigidity, and spasm of the paraspinal muscles. Rapid progression of symptoms may occur and portrays a poor prognosis. Weakness generally is asymmetric, and quadriplegia is common in patients under 1 year of age. CSF analysis shows slightly elevated proteins and mononuclear cells; analysis for viral DNA in the CSF makes the diagnosis fast and reliable. Histologically, there is parenchymal and perivascular inflammation, gliosis, and destruction of the anterior horn cells leading to atrophy. Treatment is supportive.

Fig. 8 Neuromyelitis optica in a 16-year-old girl. (a) Sagittal T1-weighted image and (b) sagittal T2-weighted image show the cervical cord is slightly swollen and displays a central cavitation (arrows, a, b) at the C3–C4 level, surrounded by edema (arrowhead, b). (c) Gd-enhanced sagittal and (d) axial T1-weighted images show enhancement of the right lateral funiculus of the spinal cord (arrow, c; arrowhead, d). (e) Coronal STIR image shows swollen, hyperintense right optic nerve (arrow), consistent with optic neuritis.
MRI is the imaging method of choice when poliomyelitis is suspected. The spinal cord shows increased T2 signal intensity involving the ventral gray matter horns (Malzberg et al. 1993). The cord may be mildly expanded at that level. The lesions enhance, and the anterior roots of the cauda equina may also enhance (Fig. 11). Similar findings to those described for the lumbar region may be seen elsewhere in the spine. The brainstem may also be involved, and this involvement may be documented by MRI (Wasserstrom et al. 1992).

Herpes Zoster Myelitis secondary to herpes zoster infection has also been reported (Friedman 1992). The symptoms and MRI findings generally correspond closely with the dermatomal distribution of the lesions. The presence of a sensory abnormality accompanied by the characteristic vesicular rash should make one suspect herpes zoster myelitis. Imaging characteristics are fairly typical: in T2-weighted images, the spinal cord shows a focal, somewhat rounded, hyperintense lesion involving one half of the

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**Table 2** Organisms more frequently involved in spinal cord infections

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<td>Streptococci</td>
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<td>Staphylococci</td>
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<td>Mycobacteria</td>
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<td>Toxoplasma</td>
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<td>Listeria</td>
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<td>Viruses</td>
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<td>Schistosoma</td>
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cord ipsilateral to the cutaneous rash (Fig. 12). Contrast enhancement may occur. It is not clear whether the myelitis is due to an allergic reaction, autoimmune vasculitis, or demyelination or as a result of direct viral infection. Viruses are generally absent in the CSF.

**Cytomegalovirus and Herpes Simplex Type I** Other herpesviruses, such as cytomegalovirus (CMV) and herpes simplex type I, may rarely involve the spinal cord and the nerve roots. MRI with gadolinium shows contrast enhancement in the posterior aspect of the cord, particularly in the lower thoracic segment. The enhancing lesion extends to the dura occasionally. The surface of the spinal cord may show a somewhat nodular pattern of enhancement. In T2-weighted images, the cord shows segments of hyperintensity. The nerve roots of the cauda equina may enhance (Fig. 13).
Human Immunodeficiency Virus  AIDS patients may present with a myelopathy (Rovira et al. 1991). This myelopathy is thought to be a direct injury by the human immunodeficiency virus. Histologically, it results in vacuolar degeneration. There is demyelination of the posterior and lateral columns. On MRI,
there is high T2 signal in the regions of myelopathy; these areas may enhance after gadolinium is given. The findings are nonspecific and seldom encountered in children.
Enterovirus 71 Enterovirus 71 (EV71) infection is an emerging epidemic disease associated with childhood acute flaccid paralysis. The most typical spinal MRI findings are those of unilateral involvement of the anterior horn cells of the spinal cord and ventral roots, appearing as hyperintense lesions on T2-weighted images. Following contrast administration, enhancement of the ventral roots, sometimes associated with enhancement of the anterior horn cells, may be seen. Bilateral anterior horn abnormalities have been associated with poor outcome (Chen et al. 2001).

Parasites
Among parasitic infections, cysticercosis is the most common form affecting the central nervous system (Mohanty et al. 1997; Chang et al. 1991). Involvement of the spinal cord occurs in about 5% of patients with cerebral cysticercosis (Castillo et al. 1988) and is very rare in children.

Most cysticercosis in the spine actually occurs within the subarachnoid spaces. These lesions are cystic with or without areas of enhancement similar to those found in the intracranial cisterns. Most parasites lodge in the distal thecal sac. Classically, the cysts are known to move according to the position of the patient. Since myelography is no longer commonly employed in these patients, this feature is seldom seen. Most intramedullary spinal cord lesions are either cystic or cystic with a peripheral nodule of contrast enhancement or appear as nonspecific ring-enhancing lesions which may also have surrounding edema.

In some cases of spinal cord cysticercosis, surgery may be indicated. Surgery may be reserved for patients with severe neurological deficits. Myelotomy with delivery of the cysts is performed. With surgery, even patients with paraplegia may have a favorable outcome.

Disorder Affecting the Nerve Roots and the Meninges

Bacterial Meningitis
Bacterial meningitis is an infectious process involving the dura, leptomeninges, and CSF. Although it is the most common infectious spinal disorder in children, imaging studies are usually not required, and the condition is diagnosed and treated on a clinical and physical examination basis. Infectious agents may enter the CNS through hematogenous spread, direct implantation (usually traumatic), local extension (secondary to sinusitis, mastoiditis, otitis, brain abscesses), and spread along the peripheral nervous system. Etiologies vary with patient age. In neonates, Streptococcus group B infections account for nearly 50% of cases, followed by E. coli and Listeria. In young infants, Haemophilus influenzae accounts for about 40–60% of cases, followed by Neisseria meningitidis and Pneumococcus. In older children and adults, Pneumococcus, Neisseria meningitidis, and Staphylococcus are the main causative agents. Tuberculosis (TB) is also increasing in the Western countries mainly owing to immigration.

Imaging Findings
The hallmark of acute-stage bacterial meningitis is infiltration of the arachnoid with inflammatory cells. In this stage, a purulent exudate diffusely covers the surface of the brain and spinal cord. This results in a diffuse enhancement of the surface of the spinal cord and nerve roots on contrast-enhanced MRI which may or may not be associated with a diffuse increase in signal intensity of the subarachnoid spaces in the T1-weighted and FLAIR images (Fig. 14). Imaging studies are usually reserved for patients suspected of harboring complications from the disease, rather than for establishment of the diagnosis. However, neuroimaging has allowed early and precise etiologic diagnosis, monitoring of treatment, and identification of complications, thereby resulting in decreased morbidity and mortality. On imaging, differential diagnosis is basically with diffuse leptomeningeal carcinomatosis (the so-called neoplastic
leptomeningitis) and with primary diffuse leptomeningeal glioneuronal tumors, which may give an almost
identical picture of thickened, diffusely enhancing pial surface of the spinal cord and nerve roots
(Gardiman et al. 2010).

An important pitfall is the postcontrast enhancement of the spinal veins, which may result in an
apparent faint enhancement of the pial surface of the cord simulating a leptomeningitis, especially at
the level of the lower thoracic cord and conus medullaris. In doubtful cases, axial images will clear the
view, showing the enhancing veins in their expected location (one anterior and two paired posterolateral
veins) (Fig. 15).

**Guillain–Barré Syndrome**

Guillain–Barré syndrome (GBS) is an acute inflammatory disorder involving the spinal and peripheral
nerves (Sladky and Ashwal 1999; Thomas and Landon 1997) and is subdivided into subtypes that are
primarily demyelinating (acute inflammatory demyelinating polyneuropathy, AIDP) or axonal (acute
motor and motor–sensory axonal neuropathy, AMAN and AMSAN) (van den Berg et al. 2014; Ryan
2013). One of the critical steps in GBS pathogenesis is the generation of antibodies that cross-react with
specific gangliosides that are present in human peripheral nerves (van den Berg et al. 2014). Specific nerve
antigens have been implicated in a number of subtypes of GBS and help in the categorization of the
syndrome; for instance, AMAN after *Campylobacter jejuni* infection is usually mediated by antibodies to
the GM1 ganglioside (van den Berg et al. 2014; Yuki et al. 2000). The Miller Fisher syndrome and
Bickerstaff brainstem encephalitis, characterized by a combination of ataxia, ophthalmoplegia, and
areflexia and considered to be cranial correlates of GBS, are characterized by anti-GQ1b antibodies

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**Fig. 14** TB meningitis in a 1.5-year-old boy. (a) Sagittal T2-weighted image is unremarkable. (b) Sagittal and (c) axial
Gd-enhanced T1-weighted images show diffuse enhancement of the subarachnoid spaces, consistent with meningitis. There
was also intracranial basal meningitis (not shown).
A few patients develop GBS shortly after receiving a vaccination, especially those against influenza; however, the risk of developing GBS after influenza infection is estimated to be four to seven times higher than after influenza vaccination (van den Berg et al. 2014).

GBS usually occurs in children, especially males; 2/3 of cases affect children between 4 and 12 years of age, but occasional neonatal cases have been described, and GBS should be included in the differential diagnosis of the “floppy infant” (Ryan 2013). GBS is presumably instigated by a prior viral disease, and such a prodromal illness, usually a respiratory illness or gastroenteritis within 2 weeks before onset, may be identified in about 65 % of patients. Clinically, it is characterized by acute onset of lower extremity weakness, progressing to flaccid paralysis and possibly ascending to involve the upper limbs, diaphragm, and cranial nerves with resulting facial paralysis and ophthalmoplegia. Sensory disturbances may be present in greater than 40 % of cases and are represented by pain (perhaps the earliest clinical symptom) and paresthesia. Sphincter dysfunction is occasionally seen, but is less common than in cases of myelitis, tumor, or other spinal cord lesions. Paralysis of the respiratory muscles is a common complication (Table 3). Ataxia is also common and results from weakness and sensory loss rather than cerebellar involvement. Younger children are especially difficult to assess and the diagnosis can be delayed in this group. Pain, difficulty with walking, and refusing to walk are the most frequent presenting symptoms in preschool children with GBS (van den Berg et al. 2014).

Table 3 Clinical manifestations of Guillain–Barré syndrome

<table>
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<tr>
<th>Onset of disease at about 7 years of age</th>
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<tr>
<td>Slightly more common in boys</td>
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<tr>
<td>Weakness in nearly 75 % of patients</td>
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<td>Pain in 55 % of patients</td>
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<tr>
<td>Ataxia in 44 % of patients</td>
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<tr>
<td>Paresthesias in 20 % of patients</td>
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<tr>
<td>Shortness of breath is rare and may indicate bulbar involvement</td>
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(Shahrizaila and Yuki 2013). Fig. 15 Vascular injection simulating leptomeningeal enhancement in a 3-year-old boy. (a) Gd-enhanced sagittal T1-weighted image shows apparent enhancement of the pial surfaces of the conus medullaris (arrows), simulating leptomeningitis. (b) Gd-enhanced axial T1-weighted images reveal a dot-like enhancement corresponding to the anterior and bilateral posterolateral spinal veins (arrows).
Histologically, there is demyelination with acute mononuclear cells infiltration, and the nerves become thick and swollen. CSF analysis shows elevation of proteins during the initial part of the disease, reflecting nerve root demyelination and a lack of inflammatory cells. Increase in the CSF protein may not be seen in the first 48 h of the onset and occasionally for as long as a week (Bradshaw and Jones 1992). GBS progresses rapidly, reaching a maximal deficit within 4 weeks. Approximately 40 % of children become non-ambulant during their illness, and up to 20 % require ventilatory support (Ryan 2013). Improvement occurs over a period of 2–18 months, and eventually greater than 90 % of patients will fully recover (Ryan 2013). Plasmapheresis is effective in adults when performed early in the course of the disease; favorable results have been reported also in children (Smith and Ouvrier 1999). Intravenous immunoglobulins are considered to be an effective and safe treatment (Smith and Ouvrier 1999). If the weakness becomes progressive and lasts for more than 2 months, the patients are said to have a chronic inflammatory demyelinating polyneuropathy (CIDP, see below).

**Imaging Findings**

MRI findings reflect the pathology of the disease (Georgy et al. 1994). After gadolinium administration, there is enhancement predominantly of the anterior nerve roots of the cauda equina (Fig. 16). Although the nerve roots may be mildly thickened, they do not display hyperintensity on T2-weighted images. Therefore, unenhanced studies are usually inconclusive (Fig. 16). Enhancement of posterior nerve roots may also occur, to the extent that in some cases there will be global thickening and enhancement of the whole cauda equina (Fig. 17). Posterior nerve root involvement may initially prevail, especially when pain predominates (Fig. 18). It should be underlined that in the very early stage of disease enhancement may be mild, if not thoroughly absent. Usually, progression to global enhancement of both anterior and posterior nerve roots occurs in a matter of a few days. On occasion, the anterior gray matter horns in the distal spinal cord will also show contrast enhancement and hyperintensity on T2-weighted imaging. Differential diagnosis is with other conditions where the blood–nerve barrier is damaged, including hereditary polyneuropathies (i.e., Charcot–Marie–Tooth and Dejerine–Sottas diseases), metabolic diseases (i.e., metachromatic leukodystrophy, Krabbe disease), neoplasia (i.e., leptomeningeal carcinomatosis, leukemia/lymphoma), and toxic exposure (i.e., methotrexate).

Involvement of cranial nerves in the same inflammatory process is called Miller Fisher syndrome (Urushutani et al. 1995). Affected patients complain with ophthalmoplegia, ptosis, facial weakness, and ataxia. MRI shows enhancement of multiple cranial nerves (Fig. 19). Differential diagnosis is with neuroborreliosis (Lyme disease).

**Chronic Inflammatory Demyelinating Polyneuropathy**

Chronic inflammatory demyelinating polyneuropathy (CIDP) is an acquired immune-mediated inflammatory disorder of the peripheral nervous system, related to GBS of which it is considered the chronic variant. Like GBS, CIDP is characterized histologically by a loss of integrity of the peripheral myelin sheath. In CDP, time to nadir of the neurological findings is longer than in GBS, usually up to 8 weeks. The course of the disease can be progressive, stepwise, or fluctuating.

Imaging studies of patients with CIDP show intrathecal as well as intra- and extraforaminal nerve root enhancement. Thus, patients with CIDP cannot be confidently discriminated from those with GBS on a single imaging study (Fig. 20). However, unlike GBS, contrast enhancement of affected nerve roots persists for months and even years after the initial diagnosis and despite treatment (Rossi et al. 2013).

**Hereditary Polyneuropathies**

Hereditary neuropathies may occur as isolated forms, in association with CNS involvement or as part of multisystem disorders (Thomas and Landon 1997).
Hereditary Motor and Sensory Neuropathies

The most common degenerative disorders of the peripheral nervous system in childhood are represented by hereditary motor and sensory neuropathies (HMSNs) (Transverse Myelitis Consortium Working Group 2002; Thomas and Landon 1997) (Tables 4 and 5).

Fig. 16 Guillain–Barré syndrome in a 2-year-old girl presenting with rapidly progressive paraparesis. (a) Sagittal T1-weighted and (b) sagittal T2-weighted images are unremarkable. (c) Gd-enhanced sagittal T1-weighted image shows enhancement of nerve roots of the cauda equina (arrows). (d) Axial T2-weighted image shows the caudal nerve roots are not thickened. (e) Gd-enhanced axial T1-weighted image shows enhancement of anterior nerve roots in the cauda equina (arrows). The diagnosis would be missed without contrast material administration.
HMSN Type I

HMSN type I is genetically heterogeneous, usually showing a dominant inheritance, although autosomal recessive and X-linked cases have been reported. It is generally referred as Charcot–Marie–Tooth disease type 1 (CMT 1). Most cases (CMT 1A) are due to duplication within band 17p11.2, which includes the gene encoding peripheral myelin protein 22 (PMP-22). CMT 1B is related to mutations of the P0 gene that maps to chromosome 1q21–23, while CMT 1C shows no linkage to chromosome 1 or 17. X-linked Charcot–Marie–Tooth disease (CMT1X) is caused by mutations in the GJB1 gene, which encodes connexin 32 (Cx32), a gap junction protein. This gene is expressed in myelinating Schwann cells.

The clinical onset of CMT 1 is usually in the first decade of life and is characterized by foot deformity or gait disturbances. Disease progression is slow. Spinal deformities will develop in about 10% of patients. Pathologically, it is characterized by extensive segmental demyelination–remyelination with the

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**Fig. 17** Guillain–Barré syndrome in a 4-year-old girl. (a–c) Gd-enhanced sagittal T1-weighted images and (d–f) Gd-enhanced axial T1-weighted images show global thickening and enhancement of both anterior and posterior nerve roots of the cauda equina.
**Fig. 18** Guillain–Barré syndrome in a 1-year-old boy presenting with paraparesis and pain. (a–c) Gd-enhanced sagittal T1-weighted images and (d–f) Gd-enhanced axial T1-weighted images show prevailing enhancement of the posterior nerve root groups (arrows).

**Fig. 19** Miller Fisher syndrome in a 7-year-old boy. (a–c) Gd-enhanced axial T1-weighted images show symmetric enhancement of the facial nerves (arrows), abducens nerves (arrowheads), trigeminal nerves (thick arrows), and oculomotor nerves (open arrows). This patient also had diffuse enhancement of the caudal nerve roots (not shown).
Fig. 20 Chronic demyelinating polyradiculoneuropathy in a 4-year-old girl. (a) Sagittal T2-weighted image is essentially normal, although some degree of nerve root clumping is suggested posterior to the conus apex (arrowhead). (b) Postcontrast sagittal T1-weighted image and (c, d) postcontrast axial T1-weighted images show diffusely thickened, enhancing caudal nerve roots (arrows). This picture is similar to that of Guillain–Barré syndrome. (e) Sagittal T2-weighted image obtained after 1 year does not reveal abnormalities; however, (f) Gd-enhanced sagittal T1-weighted image and (g, h) Gd-enhanced axial T1-weighted images show persistent, although markedly reduced, enhancement of some caudal nerve roots, consistent with a chronicized inflammatory process.
development of “onion bulbs” around the nerve fibers, degeneration of the posterior columns, loss of anterior horn cells, and degeneration of the anterior and posterior spinal roots. MRI shows significant thickening of the nerve roots in both their intradural and extradural segments (Fig. 21) (Castillo and Mukherji 1996). This may be seen in the cervical, thoracic, and lumbar regions. The thick nerve roots usually show no contrast enhancement (Castillo and Mukherji 1996). However, enhancement of hypertrophic spinal nerve roots and ganglia has been reported in a case of CMT 1 with atypical manifestations, such as progressive bladder dysfunction and severe low back pain (Fig. 22) (Cellerini et al. 2000). The dorsal root ganglia will also eventually become thick. Spinal cord impingement from enlarged intradural roots has been reported (Cellerini et al. 2000; Butesch et al. 1999). In CMT1X, brain MRI may show transient white matter changes indicative of myelin edema (Fig. 23), appearing both with and without overt symptoms including dysarthria, ataxia, hemiparesis, and tetraparesis resembling periodic paralysis and typically in association with triggering events such as fever, viral illness, and travel to high altitude (McKinney et al. 2014).

HMSN Type II HMSN type II corresponds to the classic description of CMT disease (Castillo and Mukherji 1996). It usually is inherited as an autosomal dominant trait, although autosomal recessive forms are known. The clinical picture is similar to that of HMSN type I with a later onset (during the second or third decade) and a slower course. Contrary to the type I disease, axonal degeneration is the main pathological finding and no “onion bulbs” are present. There are no specific imaging findings of HMSN type II.

### Table 4 Classification of the hereditary motor and sensory neuropathies (HMSNs)

<table>
<thead>
<tr>
<th>HMSN type</th>
<th>Pathological findings</th>
<th>Imaging findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I: hypertrophic Charcot–Marie–Tooth (CMT)</td>
<td>Segmental demyelination, few “onion bulbs,” few enlarged nerve roots</td>
<td>Occasional enlarged nerves roots of the cauda equina</td>
</tr>
<tr>
<td>Type II: neuronal Charcot–Marie–Tooth</td>
<td>Segmental demyelination, no “onion bulbs,” no enlarged nerve roots</td>
<td>No specific imaging findings</td>
</tr>
<tr>
<td>Type III: Dejerine–Sottas</td>
<td>Segmental demyelination, many “onion bulbs,” many enlarged nerve roots</td>
<td>Many enlarged peripheral or spinal nerves roots, enlarged cranial nerves</td>
</tr>
</tbody>
</table>

From Maki et al. (1999), modified

### Table 5 HMSN types I, II, and III: pathological and imaging findings

<table>
<thead>
<tr>
<th>HMSN type</th>
<th>Pathological findings</th>
<th>Imaging findings</th>
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<tbody>
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<tr>
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<td>Segmental demyelination, no “onion bulbs,” no enlarged nerve roots</td>
<td>No specific imaging findings</td>
</tr>
<tr>
<td>Type III: Dejerine–Sottas</td>
<td>Segmental demyelination, many “onion bulbs,” many enlarged nerve roots</td>
<td>Many enlarged peripheral or spinal nerves roots, enlarged cranial nerves</td>
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</table>

From Maki et al. (1999), modified
HMSN Type III HMSN type III, also known as Dejerine–Sottas disease (DSD) (Castillo and Mukherji 1996), is a highly heterogeneous group of neuropathies with variable age of onset and clinical severity. The most typical form is characterized by hypotonia and slow motor development presenting during the first year of life. Pathological findings are similar to those of HMSN type I, but more severe. The congenital forms of hereditary neuropathies, including hypomyelinating neuropathies with “onion bulbs,” are usually considered as HMSN type III forms. Approximately 15 % of cases of DSD show cranial nerve involvement (Maki et al. 1999).

Imaging findings are similar to those described above for HMSN type I. Abnormal thickening and clumping of the spinal nerve roots of the cauda equina, either with normal signal intensity (Maki
et al. 1999) or showing foci of hyperintensity in T2-weighted images, probably due to edema or demyelination (Maki et al. 1999), have been reported. Diffuse enhancement of the cauda equina nerve roots in the absence of any abnormalities on precontrast MRI has been reported in a patient with the congenital form of DSD (Fig. 24) (Cellerini et al. 2000). Spinal cord impingement and compression from enlarged intradural roots have been reported as well (Cellerini et al. 2000; Maki et al. 1999).

**HMSN Type IV** The formerly used term HMSN type IV corresponds to Refsum disease and is usually not included among the HMSNs. It is a peroxisomal disorder due to an abnormal accumulation of phytanic acid due to phytanic acid oxidase deficiency. Spinal imaging findings are nonspecific, subtle, and similar to those previously described for HMSN I.

**Fig. 22** Charcot–Marie– Tooth disease in a 10-year-old girl. (a) and (b) Gd-enhanced sagittal T1-weighted images show enhancement of caudal nerve roots (arrows), while (c) sagittal T2-weighted image is normal. (d) Gd-enhanced axial T1-weighted images confirm mild degree of enhancement that prevails in the anterior nerve roots (arrows)
Fig. 23 Charcot-Marie-Tooth disease: brain findings in a 12-year-old boy. (a) Diffusion-weighted images, (b) ADC maps, (c) axial T2-weighted images, and (d) axial FLAIR images at presentation reveal abnormal intensity across the corpus callosum (thin arrows) and centrum semiovale bilaterally (empty arrows), showing restricted diffusion. There is a posteroanterior gradient of severity. The subcortical white matter is spared. (e) Axial T2-weighted images obtained after 1 month show almost complete normalization of the findings with only a very faint residual hyperintensity in the centrum semiovale posteriorly (empty arrows)
Hereditary Sensory and Autonomic Neuropathies

The hereditary sensory and autonomic neuropathies (HSANs) are a genetically heterogeneous group of disorders pathologically characterized by degeneration of dorsal roots and ganglia. Signal intensity changes reflecting this degeneration may be demonstrated by MRI.

**Fig. 24** Lumbosacral spine in a patient with the congenital hypomyelinating form of Dejerine–Sottas disease. (a) Sagittal T1-weighted image; (b) Gd-enhanced sagittal T1-weighted image; (c) Gd-enhanced, fat-suppressed coronal T1-weighted image; (d) Gd-enhanced coronal T1-weighted image. Marked, diffuse enhancement of the cauda equina nerve roots (b) in the absence of root enlargement (a, b). Fat-suppressed image (e) enables better contrast between enhanced spinal ganglia and surrounding fat-suppressed fat tissue signal compared with corresponding non-fat-suppressed image (d) (Case courtesy of Dr. Martino Cellerini, Bergamo, Italy; reproduced with permission from Cellerini et al. (2000))
Metabolic and Degenerative CNS Disorders

A peripheral neuropathy may occur in several metabolic and degenerative CNS disorders (Table 6). Thickened, enhancing cranial nerves and caudal nerve roots are typically seen in metachromatic leukodystrophy (Fig. 25) and globoid cell leukodystrophy (Krabbe disease), and they may represent the earliest imaging finding in this context (Morana et al. 2009).

Spinal Arachnoiditis

Spinal arachnoiditis may follow infections or aseptic inflammatory processes. It is unusual in children and affects mostly males. Most childhood cases of spinal arachnoiditis are idiopathic but an antecedent of prior infectious meningitis, tuberculosis, trauma, neurofibromatosis type 1, irradiation, and syringomyelia may be elicited in some patients (Snyder 1994; Sharma et al. 1997). The symptoms include spotty or localized pain, migratory paresthesias, weakness which may also be progressive, eventual development of

Table 6  Metabolic and degenerative CNS disorders in which a peripheral neuropathy may occur

<table>
<thead>
<tr>
<th>Lysosomal disorders</th>
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<tbody>
<tr>
<td>Metachromatic leukodystrophy</td>
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<td>Krabbe leukodystrophy</td>
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<tr>
<td>Mucopolysaccharidoses</td>
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<td>Oligosaccharidoses</td>
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<tr>
<td>Fabry’s disease</td>
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<td>Farber’s disease</td>
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<tr>
<td>Gaucher’s disease</td>
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<tr>
<td>Niemann–Pick disease</td>
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<td>GM2 gangliosidosis</td>
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<th>Lipoprotein deficiencies</th>
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<tr>
<td>Bassen–Kornzweig disease</td>
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<td>Hypobetalipoproteinemia</td>
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<td>Tangier disease</td>
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<th>Disorders with defective DNA repair</th>
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<tr>
<td>Cockayne syndrome</td>
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<td>Ataxia-telangiectasia</td>
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<td>Xeroderma pigmentosum</td>
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<th>Peroxisomal disorders</th>
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<tr>
<td>Adrenomyeloneuropathy</td>
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<td>Refsum disease</td>
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<th>Mitochondrial disorders</th>
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<tbody>
<tr>
<td>NARP (neuropathy, ataxia, and retinitis pigmentosa), Leigh syndrome</td>
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<tr>
<td>Congenital disorder of glycosylation (CDG syndrome) type Ia</td>
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<tr>
<td>Cerebrotendinous xanthomatosis</td>
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<td>Chediak–Higashi disease</td>
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<td>Lowe syndrome</td>
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<tr>
<td>Vitamin E deficiency</td>
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<td>Vitamin B1, B6, and B12 and folate deficiency</td>
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<tr>
<td>Amyloidosis</td>
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<td>Porphyria</td>
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From Castillo and Mukherji (1996), Butefisch et al. (1999), modified
spastic paraparesis, and rectal and urinary sphincter dysfunction and are secondary to adhesions in the cord and nerve roots. These adhesions may also lead to the formation of CSF-containing cysts in the subarachnoid space, which may compress adjacent neural structures. Adhesions may also compromise the vascular supply to the spinal cord and nerve roots. Constant traction from spine movement makes the symptoms worse. Common symptoms are pain and paresthesias. The symptoms usually are progressive, but occasionally resolve spontaneously. Anti-inflammatory drugs and steroids

**Fig. 25** Metachromatic leukodystrophy in a 3-year-old girl. (a) Sagittal T2-weighted image is unremarkable. (b, c) Gd-enhanced sagittal T1-weighted images and (d) Gd-enhanced axial T1-weighted image show diffuse enhancement of caudal nerve roots (arrows). (e) Axial T2-weighted image and (f) axial FLAIR image of the brain show leukodystrophy with typical tigroid pattern and sparing of subcortical fibers. Similar spinal findings are also seen in Krabbe disease
usually are used to treat arachnoiditis. This treatment, however, is not satisfactory in most patients. Surgery generally is not indicated. Arachnoiditis may occasionally lead to the formation of syringomyelia.

**Imaging Findings**

The diagnosis of arachnoiditis was once obtained by myelography (Snyder 1994). However, this invasive method has now been largely surpassed by MRI with myelographic sequences. The findings are clumping and thickening of the nerve roots, adherence of the roots to the walls of the thecal sac (empty sac sign), loss of the normal configuration (blunting) of the nerve roots sleeves and distal theca, and occasionally the formation of a mass (fibrosing or ossifying arachnoiditis). Mild degrees of arachnoiditis are difficult to identify on MRI (Sharma et al. 1997). Signs of moderate to severe arachnoiditis include (Fig. 26) clumping of the nerve roots, empty thecal sac sign, enhancement of the nerve roots, and rarely an intradural mass of low T2 signal intensity which may also enhance after gadolinium administration.

Tuberculosis is a rare cause of arachnoiditis. Patients with tubercular arachnoiditis show increased T1 signal in the CSF, loss of the cord–CSF interface in all sequences, a “shaggy” cord appearance, intramedullary high T2 signal intensity, meningeal enhancement, clumping of the nerve roots of the cauda equina, nerve root enhancement, and spinal cord enhancement.

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**Fig. 26** Spinal arachnoiditis. (a) Sagittal and (b) axial T2-weighted images in a 3-year-old girl with prior surgery for spinal cord detethering show, other than a low-positioned spinal cord, clumping of the caudal nerve roots (arrowheads) within the thecal sac. (c) Axial T2-weighted image in another patient shows the lumbar and sacral nerve roots (arrowheads) are adhered to the walls of the thecal sac giving the appearance of an “empty sac”
Disorders Predominantly Affecting the Vertebra, Discs, and Epidural Space

Inflammatory Diseases

Juvenile Idiopathic Arthritis
Arthritis is a heterogeneous group of musculoskeletal disorders, comprising a large host of different diseases or conditions that affect the joints, bones, muscles, cartilage, and other connective tissues, hampering or halting physical movement. The term juvenile arthritis (JA) has a generic connotation and is used to describe autoimmune and inflammatory conditions that can develop in children aged 16 and younger. The most common form of JA is juvenile idiopathic arthritis (JIA). JIA is generally diagnosed before 16 years of age, particularly when joint symptoms persist for more than 6 weeks and an underlying infection has been excluded (Tucker et al. 1994). JIA is more common in girls than in boys and can be classified into five main types:

• Systemic arthritis, also called Still’s disease, can affect the entire body or involve many systems of the body including the internal organs and is usually associated with high fever and a rash.
• Oligoarthritis affects fewer than five joints (most commonly the knee, ankle, and wrist) in the first 6 months of the onset and can also cause uveitis or iridocyclitis.
• Polyarthritis involves five or more joints in the first 6 months of the disease and can affect the jaw, neck, hands, and feet.
• Psoriatic arthritis affects children who have both arthritis and psoriasis.
• Enthesitis-related arthritis involves the spine, hips, eyes, and entheses; it occurs mainly in boys older than 8 years with a family history of ankylosing spondylitis.

In the spine, JIA most commonly involves the cervical region. It is primarily a synovitis, but results in secondary alteration of the vertebrae. Symptoms of spine involvement are evident in about 60% of patients with JIA, whereas imaging abnormalities may be present (but clinically silent) in up to 80% of cases. The earliest signs of spinal involvement occur in the cervical region and include pain, stiffness, torticollis, and limited range motion of the head.

Imaging Findings MRI allows the direct visualization of synovitis and joint effusion (Fig. 27), shows bone marrow edema, and detects erosions before they become visible on conventional radiographs (Hospach et al. 2014). The disease generally begins at the C2–C3 level and then extends caudally or cephalad. There can be malalignment of the articular surfaces of atlantooccipital, atlantoaxial, or cervical facet joints or of two adjacent vertebrae. Interruption of the osseous joint surface with signs of inflammation, including hyperintense signal of the interarticular space, synovia, and subjacent bone in STIR images with corresponding enhancement on postcontrast T1-weighted images is classically found (Fig. 28) (Hospach et al. 2014). The dens becomes eroded, first anteriorly and then posteriorly, and may become hypertrophic causing narrowing of the spinal canal at the craniovertebral junction with possible neurological impairment due to spinal cord compression. Bone softening and basilar invagination may also occur. Contrast-enhanced MRI may also be used to discriminate between sterile joint effusions and pannus, as the latter shows enhancement (Fig. 29) (Stiskal et al. 1995). It is important to recognize that a mild amount of fluid in the connective tissue surrounding the dens is a normal finding. Calcifications in the paraspinal ligaments is said to be typical for JIA. When the thoracic spine is involved, scoliosis, fusions, and disc disease may be found. Chronically, facet and uncovertebral joint ankylosis and vertebral body fusion may also be found (Fig. 30).
In many patients, there is little relationship between symptoms and MRI findings. According to a study by Hospach et al. (2014), MRI is recommended in case of (i) signs and symptoms of cervical spine involvement, (ii) rheumatoid factor-positive polyarthritis, and (iii) rheumatoid factor-negative polyarthritis or extended oligoarthritis with recalcitrant disease necessitating intensive therapy. Scheuermann’s Disease

Scheuermann’s disease (SD), first described by H.W. Scheuermann in 1920 as “osteochondritis deformans juvenilis dorsi” (Scheuermann 1920), is an inflammatory condition of the spine that results in kyphosis of the thoracic or thoracolumbar spine. Synonyms include vertebral epiphysitis, juvenile kyphosis, or juvenile discogenic disease. SD is a common condition, occurring in approximately 5% of the general population, with a relative preponderance in males and a pronounced tendency to recur in families. The thoracic or thoracolumbar spine is most commonly involved, with rare cases involving the lumbar and, exceptionally, the cervical spine. Patients are typically adolescents complaining of rigidity or dull back pain that is generally gradual, located over the deformity, and worst at the end of the day. While the etiology remains unknown, osteochondrosis and aseptic necrosis of the ring vertebral apophyses (vertebral epiphyseal growth plates) have been advocated to play a role, perhaps exacerbated by axial spine load such as occurring in certain sport activities (Goel et al. 2014). Imaging Findings

The diagnosis is usually made on plain X-rays, showing vertebral wedging of 5° or more on three adjacent vertebral bodies; thoracic or thoracolumbar spine kyphosis of at least 40° or 30°, respectively; end plate changes or irregularities; and disc space narrowing. Additional features include Schmorl nodes, limbus vertebrae (well-corticated osseous density, usually of the anterosuperior vertebral body corner, that occurs secondary to herniation of the nucleus pulposus through the vertebral end plate beneath the ring apophysis), and spondylolisthesis (Goel et al. 2014). MRI does not really provide

![Fig. 27 Juvenile idiopathic arthritis in an 8-year-old girl. (a) Coronal STIR, (b) axial STIR, and (c) sagittal T2-weighted images show fluid collection involving the right C1–C2 joint and extending laterally (arrowheads).](image)
significant advantages over X-rays in the diagnosis of SD, except for the absence of radiation; however, it is often performed in children or adolescents in the setting of back pain. MRI (Fig. 31) will show essentially the same findings as those described above, with the addition of a variable combination of Modic-type vertebral end plate signal changes, representative of an association of bone marrow edema and inflammation (type I), conversion of red into yellow marrow as a result of ischemia (type II), and
subchondral sclerosis (type III) (Modic et al. 1988). Despite the variably significant anterior vertebral wedging and collapse, the thecal sac and spinal cord are unaffected unless there is associated disc herniation that may cause focal compression.

Disc Space Calcification
Disc space calcification (DSC), or intervertebral disc calcification, is a poorly understood, uncommon condition. First described by Baron in 1924 (Baron 1924), it is characterized by calcification of the intervertebral disc, usually in the cervical spine. Multilevel involvement is rare, but possible. Most patients are boys aged 6–10 years who complain of local pain or torticollis. Intraspinal herniation of a calcified disc fragment is a common complication (30 % of cases) (Garg et al. 2012) and may cause neurological signs such as radiculopathy or sensorimotor signs, similar to other forms of disc herniation. The etiology of DSC is unknown, and the entity is considered idiopathic in the pediatric age group. Prior trauma or inflammation has been speculated to play a role. The natural history of the condition is self-limiting, and conservative treatment is advocated (Garg et al. 2012). Half of the patients are symptom-free within 3 weeks and over 95 % within 6 months (Dai et al. 2004).

Imaging Findings Convention X-rays may show an ovoid calcification in the intervertebral disc space. CT scan confirms the presence of the calcification in the disc space (Fig. 32) and also clearly displays any extruded disc fragment that may impinge on the thecal sac and spinal cord (Fig. 33) (Calderone et al. 2009). The vertebral end plates may appear irregular, with areas of subchondral sclerosis or Schmorl nodes which are consistent with an associated spondylitis; major vertebral collapse is very rarely seen. MRI is especially useful to depict associated herniations and the effects on the spinal cord (Fig. 33) (Garg et al. 2012). Signal changes in the adjacent vertebral end plates, with hyperintensity on STIR images and contrast enhancement — better seen with fat suppression — are also indicative of an associated inflammatory process. Complete radiological normalization is reported within 3 months of the diagnosis (Dai et al. 2004); however, in our experience the healing process may be slower and lead to formation of Schmorl nodes in the adjacent end plates (Fig. 32).
Chronic Recurrent Multifocal Osteomyelitis
Chronic recurrent multifocal osteomyelitis (CRMO) is a sterile skeletal inflammation occurring primarily in childhood and adolescence, predominantly in girls. The cause is unknown; autoimmune mechanisms and genetic susceptibility have been implicated.

The disease has a long, fluctuating course with exacerbations and remissions. Pain, rigidity, and malaise are the most common complaints. Patients respond to nonsteroidal anti-inflammatory drugs, whereas antibiotics are ineffective. The diagnosis is often one of exclusion in a patient with multiple localized skeletal lesions. The long-term prognosis is good (Falip et al. 2013). Diagnostic criteria for CRMO are summarized in Table 7.

The majority of affected children have involvement of at least one long bone during their illness, most frequently the femur, tibia, or pelvic bones (Falip et al. 2013). The spine is involved in 1/3 of cases, sometime during concurrent illness. The vertebral involvement is often multifocal and may occur simultaneously with involvement of other bones. The thoracic spine is involved most commonly. Vertebral collapse is not mandatory but, when present, it may progress to kyphosis and vertebra plana.

Imaging Findings Conventional X-rays are not sensitive in the absence of clear-cut vertebral collapse and give normal results in about 50 % of cases. On CT, the involved vertebrae show a mottled lytic–sclerotic appearance; on sagittal and coronal reformats, the end plates of the involved vertebrae are typically notched, giving an impression of a wedge-like deformation (Fig. 34). MRI shows osseous edema of the involved vertebrae, which is exquisitely depicted by STIR images, and also efficiently shows end plate collapse whenever present (Fig. 34). Spine imaging is often part of a whole body STIR examination, which will also efficiently depict involvement in other bones (Falip et al. 2013). The intervertebral discs are typically spared. On postcontrast T1-weighted images, enhancement of the involved vertebrae occurs, reflecting the inflammatory involvement; the use of fat-suppressed techniques is required for adequate sensitivity (Fig. 35). Under typical conditions, the value of imaging studies is sufficient to establish a diagnosis, and vertebral biopsy is usually not necessary (Fritz et al. 2009); when performed, it will show sterile inflammation.
Infectious Diseases

Bacterial Spondylodiscitis

It generally is assumed that in children the disc is hypervascular and that infection begins there. There is now evidence that spine infection in children generally begins in the vertebral body adjacent to the end plate in the form of microabscesses (Wenger et al. 1994). Because there are many perforating vascular channels extending into the end plate and into the disc, the infection rapidly extends into these two structures. Once the disc is involved, the infection extends again superiorly and inferiorly to affect the adjacent vertebral bodies (Fig. 36). Bacteriological data is difficult to compile, as most cases of bacterial discitis and osteomyelitis are treated empirically, based on imaging studies on abnormalities. In addition, the incidence of discitis versus osteomyelitis is rapidly changing due to early diagnosis done using MRI; in the past, osteomyelitis was more common, while nowadays discitis is more frequently diagnosed. Despite this, the etiology is thought to be bacterial, especially in cases of pediatric discitis; staphylococci and Streptococcus pneumoniae account for most cases of discitis. In children with sickle cell disease, there is an increased incidence of salmonella discitis and osteomyelitis. Regardless of these factors,
staphylococci and streptococci are the most common organisms involved. The bacteria can infect the spine by hematogenous spread (arterial or venous, via the Batson plexus), external direct inoculation (i.e., after surgery, punctures, or epidural procedures), or contiguous spread from adjacent tissues (i.e., in case of retropharyngeal abscess) (Fucs et al. 2012).

Bacterial discitis is more commonly diagnosed between 2 and 8 years of age. Although the clinical diagnosis is fairly straightforward, many patients present with nonspecific findings such as failure or refusal to walk, abdominal pain, fever, and chronic back pain. Back pain is often the predominant complaint. When the cervical spine is affected, manifestations may include dysphagia and stiff neck. A young child’s inability to express himself/herself may cause delayed diagnosis (Fucs et al. 2012; Waizy et al. 2007). The onset of discitis may be gradual and subtle, progressing over the course of 2–4 weeks. Progression to weakness and paralysis suggests the formation of an epidural abscess that compresses the spinal cord and/or nerve roots. Usually, the inflammatory tests (C-reactive protein or CRP and ESR) are significantly increased, while the leukocyte count may be moderately increased or also within normal range. The clinical differential diagnosis includes infection, Scheuermann’s disease, tuberculosis, fungal infection, spinal epidural abscess, osteoid osteoma, tumor, and vertebral plana.

Fig. 32 Disc space calcification in a 7-year-old girl. (a) Conventional X-ray of the cervical spine in lateral projection shows densities in the C6–7 intervertebral space (arrow). (b) Axial CT scan and (c) sagittal reformatted CT image show calcified clusters involving the C6–C7 intervertebral disc. The C6 vertebral body is slightly reduced in height and shows a subchondral hyperdensity (arrowhead). (d) Sagittal STIR image shows the C6–C7 disc (arrow) is slightly hypointense compared to unaffected discs; the subchondral calcification also appears hypointense (arrowhead) and stands out on a diffuse background of mild hyperintensity (empty arrow) indicative of bone marrow edema. (e, f) Gd-enhanced fat-suppressed sagittal T1-weighted images show mild, diffuse enhancement of the C6 vertebral body (open arrow, e) as well as focal enhancement of the vertebral end plates at C6–C7 (arrowheads, f), suggesting a spondylitic process. (g) Gd-enhanced fat-suppressed sagittal T1-weighted image obtained at 1-year follow-up shows residual Schmorl nodes at the C6–C7 end plates (arrowheads); abnormal enhancement is no longer visible.
Imaging Findings  In the early stages of discitis, conventional radiographs have very low sensitivity and specificity (Fucs et al. 2012). The earliest radiographic sign is loss of definition and irregularity of the vertebral end plate, which precedes a stage of narrowing and erosion of the disc space (Fig. 37); eventually, in the healing phase, sclerosis of the end plates and significant narrowing of the disc space occur (Fucs et al. 2012).

MRI is the most useful method to evaluate children suspected of harboring spinal discitis/osteomyelitis particularly in the early stages, when other types of imaging, such as conventional X-rays, do not yet reveal changes (Fucs et al. 2012; Bates 1991). Although any level of the spine can be affected, the incidence of involvement of the lumbar or lumbosacral region represents the majority of cases (75 % of

### Table 7  Diagnostic criteria for chronic recurrent multifocal osteomyelitis (CRMO)

<table>
<thead>
<tr>
<th>Criteria</th>
<th>Details</th>
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<tr>
<td>Multifocal bone lesions diagnosed clinically or radiographically</td>
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<td>A characteristic prolonged, fluctuating course with recurrent episodes</td>
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<td>of pain over several years</td>
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<tr>
<td>Sites that are atypical for infectious osteomyelitis, with frequent</td>
<td></td>
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<tr>
<td>clavicular involvement and multifocality</td>
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<tr>
<td>A radiographic picture suggesting subacute or chronic osteomyelitis</td>
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<tr>
<td>No abscess formation, fistula, or sequestra</td>
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<tr>
<td>Nonspecific histopathology and laboratory findings consistent with</td>
<td></td>
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<tr>
<td>subacute or chronic osteomyelitis</td>
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<tr>
<td>Lack of causative organism</td>
<td></td>
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<tr>
<td>Occasional association with pustulosis palmoplantaris or acne</td>
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<tr>
<td>No response to antibiotic therapy and improvement with nonsteroidal</td>
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<td>anti-inflammatory drugs</td>
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From Falip et al. (2013)
patients), and the L3–4 and L4–5 interspaces are predominantly affected. In the very early stages, imaging studies are consistent with a picture of pure discitis, showing a reduction in the height of the intervertebral disc associated with swelling of the annulus, which appears hyperintense in T2-weighted images. Enhancement after gadolinium administration is evident (Fig. 37). Signal changes of the vertebral end plates and subchondral regions are initially very subtle and are more easily picked up by STIR sequences. DWI also is useful to show restricted diffusion in the involved disc (Fig. 37). As the disease progresses, the end plates may be irregular and not seen well, especially on the T1-weighted images. With advancing disease, the end plates and vertebrae become bright in T2-weighted sequences. All of these abnormalities may show gadolinium enhancement. Later on, infection may involve the paravertebral tissues (Fig. 38) or spread into other vertebral bodies via the venous plexus. In this situation, the adjacent vertebrae show abnormal signal intensity in the region of the canal for the basivertebral vein. The MRI findings lag behind clinical improvement due to adequate antibiotic treatment. When following these patients with MRI, the most important feature that predicts recovery is the absence of progression. The abnormalities may remain stable or improve slightly with the passing of time. Progression of disease indicates a failure of medical treatment.

Fig. 34 Chronic recurrent multifocal osteomyelitis in a 6-year-old boy. (a) Sagittal reformatted CT image and (b) axial CT scan show notched T4, T9, and T10 superior vertebral end plates (arrowheads) with resulting mild vertebral collapse. (c) Sagittal STIR image shows hyperintense signal from the same vertebral bodies, consistent with marrow edema
In immunocompromised patients, fungal or parasitic spondylodiscitis may be encountered (Fig. 39); in our experience, multiple disc/end plate complexes may be involved, however with a less florid picture than in immunocompetent patients.

Fig. 35 Chronic recurrent multifocal osteomyelitis in a 12-year-old girl. (a) Coronal and (d) sagittal STIR images show multiple ill-defined areas of abnormal signal intensity involving several vertebral bodies in the whole spine (arrowheads). (b) Coronal fat-suppressed T1-weighted image is inconspicuous; (e) coronal and (e) sagittal Gd-enhanced fat-suppressed T1-weighted images show enhancement of the vertebral abnormalities (arrowheads). Notice there is no vertebral collapse in this case and the intervertebral discs are spared. There is incidental hydrosyringomyelia.

Fig. 36 Proposed pathogenesis of bacterial spondylodiscitis. (a) Infection starts in the disc and propagates to the vertebral end plates. (b) Infection starts in the vertebral end plate and propagates to both the vertebral body and the disc, whence it may further propagate to the adjacent vertebra.

In immunocompromised patients, fungal or parasitic spondylodiscitis may be encountered (Fig. 39); in our experience, multiple disc/end plate complexes may be involved, however with a less florid picture than in immunocompetent patients.
Tuberculous Spondylodiscitis

Spinal tuberculosis (TB) is common in children, particularly in Third World countries, and is also increasingly seen in Western countries owing to immigration. As opposed to the adult form of the disease, childhood TB is generally more extensive and results in large abscess formation. Unlike adult TB, children seldom develop paraplegia (Ho and Leong 1994). The disease is due to *Mycobacterium tuberculosis* and infection in the chest and/or genitourinary tract precedes spinal involvement in the majority of patients.

The most frequent site for childhood spinal TB is the thoracolumbar junction (Shanley 1995). Pain and signs of chronic infection are the most typical clinical manifestations. In one series, 76% of children

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**Fig. 37** Bacterial spondylodiscitis in a 2-year-old boy presenting with refusal to walk. (a) Conventional X-ray of the lumbar spine, lateral projection, shows disc space narrowing at L4–L5. (b) Sagittal T1-weighted and (c) sagittal STIR images confirm significant disc height reduction with a blurring of the adjacent central end plates (arrowheads). The signal from the vertebral bone marrow is diffusely abnormal. (d) Gd-enhanced sagittal T1-weighted image shows collection with marginal enhancement involving the central portion of the L4–L5 disc and extending into the adjacent end plates (arrowheads); in the absence of fat suppression, enhancement of the vertebral bodies does not stand out in comparison with the unaffected bodies. (e, f) Gd-enhanced axial T1-weighted image shows discal abscess (arrowheads) extending through a cortical erosion (curved arrow) into the right paravertebral space (arrows). (g, h) Axial diffusion-weighted image shows restricted diffusion both in the disc space (arrowhead) and paravertebral region (arrow)
affected were younger than 5 years of age, nearly 50 % of children had neurological deficits on hospital admission, 50 % of patients recovered within 6 months of appropriate therapy, and paraspinal abscesses were found in 62 % of patients. Diagnosis is difficult and may be confirmed only by positive histology and/or culture. Often, the diagnosis is based on clinical manifestations, radiographic findings, and response to antibiotics. TB in the lumbosacral region is uncommon, and other etiologies such as brucellosis should be considered when this area is primarily involved (Sharif et al. 1990). Craniocervical involvement may also be seen in children and is accompanied by significant abscess formation. Cervical involvement is almost always accompanied by neighboring nodal disease. Noncontiguous involvement of multiple vertebrae, also termed skip lesions, is found in approximately 16 % of cases of spinal TB (Polley and Dunn 2009). Imaging of the entire spine is mandatory in order not to overlook distant vertebral involvement. According to the location of the infection, three patterns have been described: anterior, paradiscal, and central (Fig. 40).
Imaging Findings  In the anterior type (Fig. 41), the infection begins in the anterior (and generally also inferior) vertebral body and extends under the anterior longitudinal ligament to involve other vertebrae. In the paradiscal type (Fig. 42), the infection begins in the lateral sides of the disc and results in narrowing of the disc space. Paradiscal disease is the least common form in children. In the central type, the infection begins in the middle of the vertebral body (Fig. 43) and may extend posteriorly into the spinal canal resulting in an epidural abscess (Fig. 44). It is noteworthy that discal involvement may occur late during the course of the disease, whereas early on the disc can be completely preserved, resulting in a pure spondylitic process (Fig. 45); under such circumstances, the differential diagnosis from vertebral tumors, which also typically spare the disc space, may not be feasible based on imaging alone.

Despite antibiotic treatment, infection may rapidly progress (Fig. 46), may cause vertebral collapse (Fig. 44), and may eventually result in acute-angle kyphosis (i.e., Pott’s disease) (Fig. 47). TB may also involve the posterior elements in an isolated fashion. Before frank abscess formation there is a stage characterized by the development of masses of granulation tissues.

Epidural Abscess
An epidural, or extradural, abscess is a collection of pus between the bone and the dura mater (Wenger et al. 1994). Epidural abscesses are commonly secondary to pyogenic or tuberculous discitis (Fig. 44) and osteomyelitis (Ruiz et al. 1995). In the absence of discal and vertebral involvement, the infection usually is from hematogenous spread of primary foci in the urinary tract, skin, lungs, and teeth. Patients with AIDS and long-term vascular access catheters are also prone to develop epidural abscesses. Rarely, epidural abscesses are a complication of lumbar tap or infected epidermoids.
The most common clinical signs are pain, fever, and rapidly progressing neurological symptoms. In children, neurological signs may be absent or masked by prior administration of antibiotics. From the clinical viewpoint, the differential diagnosis mainly includes ATM, tumor, trauma, tuberculosis, and herniated disc.

Fig. 40 Tubercular spondylodiscitis: the three modes of propagation. (a) Anterior type: infection starts in the anterior vertebral body and spreads under the anterior longitudinal ligament to the adjacent vertebrae. The disc space is relatively spared until late during the course of disease. (b) Paradiscal type: infection starts in the vertebral end plate, as in bacterial spondylodiscitis. The disc space is involved in the early stages of disease. This is the rarest form in children. (c) Central type: infection starts in the central vertebral body and may propagate to adjacent vertebrae under the posterior longitudinal ligament, thereby resulting in thecal sac compression. Also in this variety, the disc space is initially relatively spared.

Fig. 41 Tubercular spondylodiscitis: anterior type in an 11-year-old boy. (a) Sagittal T2-weighted image shows narrowed, hypointense T8–9 disc space (thin arrow). Infection starts in the anterior portion of the T8 vertebra and generates a huge pre- or paravertebral abscess (arrowheads); infection also propagates to the T9 vertebral body, which is also T2 hyperintense. (b, c) Axial and (d) coronal Gd-enhanced T1-weighted images show huge paravertebral abscesses bilaterally, originating from a discontinuity of the anterior vertebral wall of T8 (arrows, b)
Epidural abscesses have a poor prognosis and high mortality. Rapid progression is generally an indication for the need of surgical decompression, and most patients will remain with deficits. Fortunately, children tend to recover in a more complete fashion than adults. Lumbar puncture is contraindicated in patients suspected of having an epidural abscess, and MRI is the diagnostic method of choice in such instances.

**Imaging Findings** Early on, MRI may detect a prominent epidural space showing intermediate signal intensity in both T1- and T2-weighted images. The abnormality enhances homogeneously after...
gadolinium administration and is most often a phlegmon. Diffusion-weighted imaging (DWI) shows restricted diffusion, with high signal on trace and low signal on ADC maps. Surgery is not indicated in these patients, as they improve considerably after appropriate antibiotic therapy. In cases of frank abscess formation, there is a rim enhancing abnormality in the epidural space (arrowhead). The L2–3 disc space is essentially uninvolved (thin arrows). Notice there is no spinal canal invasion. (d, e) Axial CT scans show central necrosis (arrowheads) with interrupted posterior body wall

Fig. 43  Tubercular spondylodiscitis: central type in a 2-year-old girl. (a) Sagittal T1-weighted image, (b) sagittal T2-weighted image, and (c) Gd-enhanced sagittal T1-weighted image show infection involving the central portion of the L3 vertebral body (arrowhead). The L2–3 disc space is essentially uninvolved (thin arrows). Notice there is no spinal canal invasion. (d, e) Axial CT scans show central necrosis (arrowheads) with interrupted posterior body wall
Fig. 44 Tubercular spondylodiscitis: central type with posterior spinal canal invasion and epidural abscess in an 8-year-old boy. (a) Sagittal T1-weighted image, (b) sagittal T2-weighted image, and (c) Gd-enhanced sagittal T1-weighted image show infection starts in the central portion of the L4 vertebral body, disrupts the posterior vertebral wall, and propagates into the spinal canal below the posterior longitudinal ligament, forming an epidural abscess (arrows). The L3–4 disc space is irregular and probably already involved, albeit without frank, diffuse enhancement. (d) Gd-enhanced axial T1-weighted image shows the thecal sac (arrow) is markedly compressed by the ventrally located collection. (e) Axial CT scan shows central-posterior vertebral body necrosis (arrowheads) with disruption of the posterior body wall. (f) Sagittal T2-weighted image obtained after 8 months, following antibiotic therapy, shows pathological tissue is completely absent; however, the L3–L4 disc is dehydrated, and marked reduction of the height of the L4 vertebral body is evident (arrowhead)
Fig. 45  Pure tubercular spondylitis in a 2-year-old boy. (a) Sagittal T1-weighted image and (b) Gd-enhanced fat-suppressed sagittal T1-weighted image show diffuse involvement of the L5 vertebral body, which appears hypointense on the baseline T1-weighted image and enhances after Gd administration. Within the central–posterior portion of the body, a necrotic lesion (arrowheads) is seen. (c) Axial T2-weighted image and (d) Gd-enhanced fat-suppressed axial T1-weighted image also show the necrotic lesion involving the right portion of the L5 vertebral body and abutting the epidural spaces posteriorly to slightly compress the thecal sac. In the absence of discal involvement, differentiation from neoplasm is not immediate.

Fig. 46  Tubercular spondylodiscitis: rapid evolution in a 4-year-old boy. (a) Sagittal T2-weighted image at presentation shows extensive abnormalities involving the L4 and L5 vertebral bodies and a reduction in height of the L4–L5 disc (arrow). (b) Sagittal CT reformatted image obtained at 2 months following antibiotic treatment shows extensive necrosis of the L4 and L5 vertebral bodies, which are almost completely replaced by a necrotic collection, with resulting significant vertebral collapse; notice the blown-out L4 anterior body wall (arrowhead). (c) Sagittal T2-weighted image shows extensive involvement of the L4 and L5 vertebral bodies, with complete disappearance of the intervening disc.
of the abscess. This finding is presumed to represent cord edema secondary to compromised venous drainage, secondary to the involvement of Batson plexus. Occasionally, arterial compromise with subsequent cord ischemia may also occur. The sensitivity of MRI for the detection of epidural abscesses is said to be over 90 %.

References


