Evaluating the child with unsteady gait

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ABSTRACT

يعتبر خلل التوازن أثناء المشي من الأعراض الشائعة لدى الأطفال بقسم الطوارئ والأعصاب. تتعدد أسباب خلل التوازن، ولكن من أهم اولويات التقييم الأولى هو التأكد من عدم وجود التهاب أوٍ ورم بالدماغ. التعرف علي المسببات الحميدة والغير عصبية أيضاً مهم لتفادي القيام بفحوصات متعددة دون الحاجة إليها أو تنويم المريض بالمستشفى. في هذه المقالة النقدية نقدم مراجعة حديثة عن تقييم الطفل المصاب بخلل التوازن مع مناقشة الفحوصات اللازمة والعلاج. قد يكوٍن خلل التوازن ناتج عن مرض بالمخيخ أو مشكلة حسية، علماً بأن أمراض المخيخ قد تكون حادة، مزمنة، متدهورة، أو متقطعة. وتتعدد أسباب هذه المشكلة و منها الإصابات، الالتهابات، أمراض الاستقلاب، العيوب الخلقية، والأورام. أما أسباب خلل التوازن الناتج عن مشاكل الإحساس فيكوٍن بسبب تأثر في الأعصاب الطرفية أو الحبل الشوكي. علماً بأن التقييم الدقيق يعتمد علي أخذ التاريخ المرضي بدقة ومن ثم الفحص السريري الذي يرشد إلي عدد ونوع الفحوصات المطلوبة للوصول إلي التشخيص الدقيق.

Unsteady gait is a relatively common presentation to the pediatric emergency and neurology services. Unsteadiness can be due to a wide variety of causes, however, the primary concern on initial assessment is to exclude serious disorders such as meningitis, encephalitis, or brain tumors. Recognizing benign and non-neurological causes of unsteady gait is essential to avoid unnecessary investigations and hospital admission. In this review, a clinical approach to the unsteady child is presented with discussion of diagnostic considerations, approach to investigation, treatment, and prognosis. Ataxia can be cerebellar or sensory. Cerebellar ataxia can be acute, chronic, progressive, or episodic. It may result from trauma, infections, metabolic, degenerative disease, space occupying lesions, or congenital anomalies. Sensory ataxia is due to peripheral neuropathy involving large myelinated fibers that carry vibration and position sense, or due to posterior spinal column dysfunction. Accurate assessment depends on detailed history, examination, and then formulation of a differential diagnosis list to guide laboratory investigations.

Neurosciences 2009; Vol. 14 (1): 3-9

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Neurological disorders are common in Saudi Arabia accounting for up to 30% of all consultations to pediatrics.[1] Trauma, ingestion, and acute neurological disorders are common, mainly as a result of improper safety practices of many parents.[2] Consanguineous marriages also add to the problem, resulting in increased prevalence of many inherited and genetic neurological disorders.[3,4] Unsteadiness and ataxia are relatively common neurological presentations of a variety of these acute and chronic disorders. Accurate assessment includes detailed history, examination, and then formulation of a differential diagnosis list to guide laboratory investigations.[5-7] Many students, residents, and generalists consider the assessment of neurological disorders one of the most difficult aspects of their clinical practice.[8-10] In this review, a clinical approach to the unsteady child is presented with discussion of diagnostic considerations, approach to investigation, treatment, and prognosis.

Unsteady gait. Gait unsteadiness is not always due to neurological causes (**Table 1**). In fact, simple injuries and musculoskeletal etiologies are most common. In the toddler, skeletal abnormalities should be suspected as they could mimic other causes of unsteady gait. It is important to recognize benign and non-neurological causes to avoid unnecessary investigations and hospitalization. Functional (hysterical) gait disorders are also not uncommon, especially in female adolescents, and should be suspected when the gait is not wide based or when falls are rare (**Table 1**). Seizure related ataxia (post-ictal, nonconvulsive status epilepticus) should be considered in children with frequent or prolonged seizures (**Table 2**). In this situation, the unsteadiness may also be related to antiepileptic drugs, such as benzodiazepines or barbiturates, and therefore should resolve slowly over a day or 2.[3]

Table 1 - Causes of unsteady gait in children.

Foot deformity

Skeletal abnormalities (ankle, knee, or hip joint)

Antalgic gait (due to pain)

Migraine (basilar migraine, benign paroxysmal vertigo)

Raised intracranial pressure (hydrocephalus)

Paretic ataxia (weakness due to upper or lower motor neuron lesion)

Cerebellar ataxia
 Congenital/genetic
 Traumatic (contusion, hemorrhage, post-concussion, vertebrobasilar dissection)
 Toxic and drugs (alcohol, antihistamines, anticonvulsants)
 Infectious/immune-mediated (chicken pox, ADEM, brainstem encephalitis, MS)
 Malignancy (medulloblastoma, neuroblastoma)
 Paraneoplastic (opsoclonus-myoclonus syndrome)
 Vascular (stroke, hypertension, AV malformation, blood disorders)
 Degenerative (ataxia telangiectasia)
 Post-ictal (epileptic ataxia)

Sensory ataxia
 Guillain-Barré syndrome, chemotherapy, heavy metals, B6, B12 deficiency

Functional ataxia (including Münchhausen by proxy syndrome)

ADEM - acute demyelinating encephalomyelitis,
MS - multiple sclerosis, AV - arteriovenous

Table 2 - Types of cerebellar ataxia in childhood.

Acute
 Trauma
 Toxic and drugs
 Seizure related (post-ictal, nonconvulsive status epilepticus)
 Infections/postinfectious
 Vascular (stroke, hypertension, AV malformation, blood disorders)
 Malignancy (medulloblastoma, neuroblastoma)
 Paraneoplastic (opsoclonus-myoclonus syndrome)
 Functional

Chronic
 Congenital (cerebellar hypoplasia, Dandy-Walker and Chiari malformation)
 Posttraumatic
 Following meningitis/encephalitis
 Post-tumor resection or radiation
 Hypoxic-ischemic insult

Progressive
 Friedreich ataxia
 Ataxia telangiectasia
 Sphingolipidosis (gangliosidosis, Niemann-Pick disease)
 Leukodystrophies (Pelizaes-Merzbacher, Krabbe, metachromatic leukodystrophy)
 Mitochondrial disorders (Leigh disease, MERF)
 Neuronal ceroid-lipofuscinosis
 Progressive myoclonic epilepsies (Lafora disease, Uverricht-Lundborg disease)
 Congenital defect of glycosylation
 Abetalipoproteinemia

Recurrent
 Migraine (basilar migraine, benign paroxysmal vertigo)
 Genetic (autosomal dominant episodic ataxias)
 Metabolic (amino acidopathies, urea cycle disorders, mitochondrial disorders)

AV - arteriovenous, MERF - myoclonic epilepsy with ragged red fibers

Ataxia. Ataxia (lacking order in Greek) refers to a pathologic abnormality of organization or modulation of movement. Although ataxia is most commonly attributable to cerebellar dysfunction, lesions at several levels of the nervous system can result in motor incoordination. Ataxia may be congenital or acquired. Congenital ataxia is usually associated with central nervous system malformations. Acquired ataxia can be classified as acute, chronic, or episodic (**Table 2**). Episodic and chronic progressive ataxias are less common in children and are usually caused by inherited metabolic or genetic disorders.[11]

Clinical evaluation. History. Most children with ataxia are seen because of refusal to walk or wide-based or drunken gait. Parents less commonly notice the involvement of the arms (tremor), head (titubation), trunk (inability to sit steadily), and speech (dysarthria). At presentation, the primary concern is to exclude serious causes of acute ataxia, including infections and tumors (**Table 1**). Detailed history will frequently clarify the cause of the unsteady gait.[12] Inquiry of prior or current symptoms of systemic infection should be included. History of trauma, infection, drug ingestion or headaches may suggest important associations (**Tables 1 & 2**). Recurrent or persistent headache and vomiting or diplopia suggests raised intracranial pressure. A common cause of acute ataxia is inadvertent or deliberate drug ingestion. The child's activities should be reviewed to explore possible exposure to medications, alcohol, and household chemicals. Keep in mind that the causes of acute ataxia are quite different from those of chronic or progressive ataxia. Acute ataxia can be related to trauma, vascular insults, infection, or drug ingestion, while chronic progressive ataxia suggests an inherited metabolic, degenerative, or neoplastic etiology (**Table 2**). Recent immunizations should be noted, as should the child's general state of health prior to presentation. Some inherited metabolic disorders, such as mitochondrial cytopathies and maple syrup urine disease, may present with intermittent ataxia that resolves slowly (**Table 3**). The child may be initially normal in-between the attacks. These episodes are frequently precipitated by infections or drug ingestion (for example, valproic acid) and result subsequently in chronic progressive sequelae. Accurate past medical and family histories are important in eliciting the possible diagnosis in these situations.

Examination. Physical examination can be difficult as ataxic children are often uncooperative and irritable.[13] General examination includes examination of the skin for neurocutaneous signs and examination for meningeal irritation. Examination of the back, hips, lower limbs, and feet is needed to exclude musculoskeletal causes. Observation for signs of trauma or arthritis is needed. Examination of the eyes may provide some clues (for

example, conjunctival telangiectasia). Nystagmus is common to disorders affecting the cerebellar hemispheres. Additional abnormalities of cranial nerve examination, such as papilledema and cranial nerve palsies, suggest a space occupying lesion or hydrocephalus. Pupillary abnormalities can be seen with mass lesions, raised intracranial pressure, stroke, or intoxication. Mental status examination in children with postinfectious cerebellar ataxia reveals normal alertness. Abrupt altered responsiveness suggests drug ingestion or toxic exposure. Extreme irritability can be seen in meningitis, encephalitis, and opsoclonus-myoclonus syndrome. Detailed motor examination is needed to exclude weakness, which may result in hypotonia and incoordination. Cerebellar ataxia is characterized by hypotonia, wide based gait, and dysarthria (fluctuations in clarity, rhythm, tone, and volume). Examination of coordination starts by examining the gait. In cerebellar disease, the patient is off balance with eyes open and worse with eye closure. Walking on a straight line will identify unilateral hemispheric cerebellar disease as the patient will sway towards the affected side. Midline (vermal) lesions cause dysarthria, truncal titubation, and gait abnormalities, whereas lesions of the cerebellar hemispheres spare speech but result in ipsilateral limb hypotonia, dysmetria, and tremor. Tandem walk (walking on a straight line with feet closely attached and alternating in front of each other) is more difficult to perform and may identify subtle cerebellar ataxia. Dysmetria (poor coordination of voluntary

Table 3 - Causes of recurrent intermittent ataxia in childhood.

Migraine and migraine variants
    Basilar migraine
    Benign paroxysmal vertigo
    Benign paroxysmal torticollis of infancy
    Alternating hemiplegia of childhood

Genetic disorders
    Episodic ataxia type 1 (paroxysmal ataxia with myokymia)
    Episodic ataxia type 2 (acetazolamide responsive)
    Episodic ataxia types 3 and 4
    Episodic ataxia with paroxysmal dystonia

Metabolic disorders
  Amino acidopathies
    Hartnup disease
    Maple syrup urine disease
  Urea cycle disorders
    Carbamoyl phosphate synthetase deficiency
    Ornithine transcarbamylase deficiency
    Arginosuccinic aciduria
  Organic acidopathies
    Biotinidase deficiency
    Isovaleric acidemia
  Mitochondrial disorders
    Pyruvate dehydrogenase deficiency
    Leigh disease
  Carnitine acetyltransferase deficiency

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movements) results in overshooting of limb movements and difficulty with rapid alternating movements (dysdiadochokinesia). Therefore, finger nose or heel shin testing, rapid alternate hand movements or foot tap will test for limb ataxia. Note that the arms have to be adequately stretched during the finger nose test to identify intention tremor as the amplitude of this tremor increases as it reaches the target. The tremor is a to and fro oscillation perpendicular to the approached object. The deep tendon reflexes can be pendular, with slowed contraction and relaxation phases. Detailed sensory examination is needed to exclude sensory ataxia. Particular attention is needed for vibration and position sense examination. Romberg's sign will help in testing position sense as the patient stands with outstretched hands and closely placed feet. Off balance with eye closure represents a positive sign, indicating sensory ataxia.

Cerebellar ataxia. Acute cerebellar ataxia. Inadvertent or deliberate drug or toxic ingestion is an important and common cause of acute ataxia. Anticonvulsants, benzodiazepines, alcohol, and antihistamines are commonly implicated agents.[14] A high index of suspicion should always be maintained as a history of ingestion or exposure might not be forthcoming. In this situation, the ataxia is often accompanied by mental status changes such as lethargy, confusion, inappropriate speech, or behavior. Postinfectious cerebellar ataxia accounts for 40% of all cases of childhood ataxia. It usually results from cerebellar demyelination, or less commonly a result of direct cerebellar infection.[15] The demyelination is an autoimmune phenomenon incited by infection or immunization, with subsequent cross-reaction of antibodies against the cerebellum. History of antecedent illness 1-3 weeks before presentation is obtained in around 70% of patients. Numerous infectious agents have been implicated in the pathogenesis of this condition. As many as 26% of cases are preceded by varicella. Rarely, the development of ataxia precedes the eruptive phase of varicella infection.[16] The introduction of universal immunization against varicella is likely to render varicella-related cerebellar ataxia uncommon. The disorder is most common in younger boys (2-4 years) but may be seen in adolescents. Postinfectious cerebellitis presents with acute gait abnormalities, ranging in severity from unsteadiness and a wide-based stance to complete inability to walk. Symptoms are maximal at onset and may be more severe in cases following varicella infection.[17] The extremities are less affected than the trunk. Acute ataxia is also a common feature of acute demyelinating encephalomyelitis (ADEM), which also develops after a viral illness or vaccination.[18] However, ADEM is distinguished from post-infectious cerebellar ataxia by the occurrence of alteration of consciousness,

Table 4 - Differentiating clinical, laboratory, and MRI features of ADEM and MS at initial presentation.[19]

Feature	ADEM	MS
History of recent infection or vaccination	common	rare
Systemic symptoms	common	rare
Fever	43%	6%
Headache	57%	24%
Fatigue	71%	29%
Vomiting	57%	0%
Encephalopathy	71%	6%
Seizures	Common	rare
Isolated optic neuritis	Less common (23%)	common
Severity of illness	Usually severe	Less severe
CSF oligoclonal bands	29%	75%
MRI changes		
Periventricular distribution	50%	91%
Involvement of the corpus callosum	17%	64%
Recurrence	Rare	Common

ADEM - acute disseminated encephalomyelitis, MS - multiple sclerosis

seizures, and multifocal neurologic deficits.[19] Systemic symptoms such as fever, headache, and meningism are also more common (**Table 4**). Repeated episodes of demyelination should raise the concern for multiple sclerosis (MS).[20] Associated findings include optic neuritis and long-tract signs. Head injuries may cause acute ataxia owing to concussion, cerebellar contusion or hemorrhage. After neck injuries, ataxia can point to vertebral artery dissection resulting in ischemic stroke.[21] Cerebellar hemorrhage is rare in childhood and, in the absence of a bleeding diathesis, is most commonly associated with arteriovenous malformations.[22] Acute ataxia is occasionally associated with rapid chaotic multidirectional conjugate eye movements (opsoclonus), myoclonus, and encephalopathy in the so called opsoclonus-myoclonus syndrome. The syndrome can be postinfectious, or the presenting manifestation of an occult neuroblastoma.[23,24] Ataxia in this disorder can be of subacute onset and can fluctuate in association with irritability and recurrent vomiting. Ataxia has also been described as a paraneoplastic phenomenon in other malignancies, such as Hodgkin's disease, histiocytosis, and hepatoblastoma.

Chronic cerebellar ataxia. Cerebellar ataxia may result from static (non-progressive) cerebellar anomalies or insults (**Table 2**). Congenital causes include cerebellar hemispheric hypoplasia (**Figure 1**), vermal aplasia, basilar impression, and Chiari malformation type 1. Cerebellar insults following trauma, infection, hypoxia, or ischemia are usually associated with other motor,

developmental, and cognitive deficits.[25] In many of these disorders, including cerebral palsy, the child is expected to make some improvements with time. This improvement is not only dependent on the severity of the underlying etiology, but also on associated motor deficits (weakness and spasticity), therapeutic interventions, and rehabilitation.[25]

Progressive cerebellar ataxia. Brain tumors are important causes of progressive cerebellar ataxia. Around 50% of all brain tumors arise from the brain stem or cerebellum.[26] Posterior fossa tumors usually present with slowly progressive ataxia and symptoms of increased intracranial pressure. Other clinical features can include headache, papilledema, personality change, and focal neurological abnormalities.[27] Occasionally, supratentorial tumors produce ataxia, usually those in the midline. Parenchymal lesions of the frontal lobes can cause ataxia due to involvement of frontocerebellar associative fibers.[28] A number of metabolic and degenerative disorders may present predominantly with cerebellar ataxia (**Table 2**). Progressive ataxia is seen in all patients with ataxia telangiectasia and spinocerebellar ataxias such as Friedreich ataxia.[29,30] Rare disorders, such as glucose transporter deficiency syndrome, should also be considered.[31] This disorder is characterized by ataxia, developmental delay, difficult epilepsy, and low CSF glucose (<2.2 mmol/L). Rapid and impressive improvement is noted after instituting the ketogenic diet.[32]

Recurrent cerebellar ataxia. Occasionally, migraine may present with intermittent ataxia. Less common

Figure 1 - An infant with incoordination and hypotonia due to congenital brain malformation (cerebellar hypoplasia).

periodic syndromes of childhood that are precursors to migraine are listed in **Table 3**. These syndromes, quite peculiar to children, present a wide variety of episodic symptoms, such as abnormal movements, vomiting, ataxia, and vertigo, and may not include headache at all.[33] Autosomal dominant episodic ataxias are related to mutations in ion channel genes. Treatment with acetazolamide or phenytoin may be helpful. Intermittent ataxia should particularly raise the suspicion of an underlying inborn error of metabolism (**Table 3**). Acute exacerbation develops after high protein ingestion, concurrent febrile illness, or other physical stress. This rare presentation occurs mainly in the late infantile and juvenile partial forms of the metabolic disorders listed in **Table 3**. Intermittent cerebellar ataxia may or may not be associated with concomitant episodes of stupor and vomiting.

Sensory ataxia. Ataxia can result from loss of sensory input to the cerebellum due to lesions in the posterior column of the spinal cord, roots, or peripheral nerves. Sensory ataxias are characterized by a positive Romberg's sign and decreased deep tendon reflexes. Loss of posterior column sensory functions (proprioception and vibration sense) causes incoordination of the hands and a wide-based, "steppage" gait. These findings worsen with the eyes closed. Sensory ataxia can be seen in children with severe peripheral neuropathy, such as Dejerine-Sottas disease. Guillain-Barré syndrome (GBS) can result in ataxia, usually in association with weakness and pain.[34] However, ataxia is a rare presenting symptom.[35] Ataxia is characteristic of the less common Miller Fisher variant of GBS, which is a triad of ataxia, areflexia, and ophthalmoplegia.[36] In this disorder, ataxia is often more marked in the extremities.

Investigations. The primary aim of investigations is to exclude serious conditions, such as brain tumors. A thorough history and physical examination are far more likely to identify the etiology of acute ataxia than is a battery of screening investigations. Of all investigations, drug screen and neuroimaging are most important. However, in the absence of altered consciousness, focal neurologic signs, or marked asymmetry of ataxia, the yield of neuroimaging is low. Posterior fossa abnormalities can be difficult to visualize on CT scans, where artifacts sometimes obscure the brain stem and cerebellum. Magnetic resonance imaging with diffusion is preferred. Acute postinfectious cerebellar ataxia is a diagnosis of exclusion. The MRI of the brain is normal in most children with postinfectious ataxia. Lesions suggestive of focal cerebellar demyelination are occasionally identified. In ADEM, MRI demonstrates multiple asymmetrical foci of demyelination that often enhance with gadolinium contrast, reflecting local breakdown of the blood-brain barrier (**Table 4**). Brainstem encephalitis

is associated with areas of increased signal intensity within the brain stem and cerebral peduncles on T2-weighted MRI.[37] Neuroimaging is usually normal at presentation of opsoclonus-myoclonus syndrome, however, some degree of cerebellar atrophy can be seen on follow-up imaging.[38] Cerebrospinal fluid (CSF) examination is commonly normal in postinfectious ataxia and ADEM, with mild pleocytosis and elevation of protein. Significant pleocytosis, low glucose, and elevation of CSF protein suggest meningitis or encephalitis. The CSF examination can demonstrate cytoalbuminologic dissociation in more than 90% of patients with GBS. However, the CSF can be normal in the first week of the disease. Oligoclonal bands and elevation of the serum:CSF immunoglobulin index and myelin basic protein level are characteristic of MS, however, they can also be present in ADEM.[19] Differentiating features between MS and ADEM are summarized in **Table 4**. Electromyography (EMG) is indicated where sensory ataxia is suspected. In GBS, EMG can be normal at the onset and only shows evidence of proximal demyelination after several days to a week of symptom onset. Urinary excretion of catecholamine metabolites is increased in up to 60% of patients with paraneoplastic opsoclonus-myoclonus syndrome.[39] A CT or MRI of the chest and abdomen is needed to identify small tumors. Investigations of children with progressive ataxia are directed towards identifying the underlying etiology and examining associated complications (for example, immune deficiency). The findings on history and physical examination will guide the physician in selecting the required laboratory investigations. For example, if ataxia-telangiectasia is suspected, serum IgA, IgE, and alpha-fetoprotein should be obtained. Serum lipid profile is abnormal in abetalipoproteinemia or hypobetalipoproteinemia. Basic blood works may prove useful in certain disorders. Complete blood count may reveal pancytopenia in certain organic acidopathies (for example, isovaleric, propionic, and methylmalonic acidemias). Blood film may show vacuolated lymphocytes in neuronal ceroid lipofuscinosis, fucosidosis, and sialidosis. Blood gas analysis will detect metabolic acidosis in many metabolic disorders such as organic acidopathies, urea cycle disorders, and mitochondrial encephalopathies. Serum ammonia, lactate, pyruvate, amino acids, and urine for amino acids and organic acids would screen for most amino acid disorders, organic acidopathies, and urea cycle abnormalities. Liver function tests are disturbed in neurovisceral sphingolipidosis and certain gray matter neurodegenerative disorders (for example, progressive infantile poliodystrophy). Chest x-ray may reveal cardiomegaly in early mitochondrial disorders and Friedreich ataxia. Neuroimaging, particularly

MRI, can show characteristic features in several neurodegenerative disorders such as leukodystrophies and Leigh disease. Frequently, specific diagnostic tests and enzyme assays are needed to reach a definitive diagnosis. These specialized tests are expensive and should be used selectively. They will frequently involve skin fibroblast culture, CSF examination, DNA studies, nerve, or muscle biopsy.

Management. The majority of causes of acute cerebellar ataxia are reversible. Treatment of ingestions depends on the nature and amount of the ingested substance. In some cases, administration of an antidote, chelation, dialysis, or other therapies is required. In post-infectious cerebellitis, the improvement usually begins after the first week and completes within 3 months.[40] There is no evidence that immunosuppressive therapy, such as steroids, improves the outcome. Recovery from ADEM is typically slower and can be hastened by treatment with corticosteroids.[41] Our protocol involves using 15 mg/kg/day of IV methylprednisolone in 4 divided doses for 3 days followed by one mg/kg/day oral prednisone as single morning dose for 2 weeks. This dose is then tapered slowly over 3-4 weeks. Most patients with ADEM recover completely; however, a minority has significant sequelae.[42] Relapses are rare and should raise the possibility of multiple sclerosis (**Table 4**). Brainstem encephalitis should be treated empirically with broad spectrum antibiotics and acyclovir.[43] Outcome in most other conditions associated with ataxia is dependent on the underlying disease process. Tumors, stroke, and traumatic brain injury are commonly complicated by significant sequelae. Neuroblastoma should be removed surgically. The underlying tumor is small and well differentiated in most cases of paraneoplastic opsoclonus-myoclonus syndrome. The neurologic syndrome of opsoclonus-myoclonus syndrome can improve slowly; however, most children have long-term neurological, cognitive, and behavioral deficits.[44] Some patients respond to high-dose steroids or intravenous immunoglobulin (IVIG). Children with GBS should be admitted to hospital, with careful monitoring of their respiratory and autonomic function.[45] Specific treatments include IVIG and plasmapheresis. Intravenous immunoglobulin is safer and should be used when children lose ambulation or develop significant respiratory or bulbar dysfunction. More than 90% of children with GBS recover completely within 3-6 months of disease onset.[46] Chronic ataxia due to static insults and cerebral palsy usually improves with time.[47] Ataxia may persist after operating on children with posterior fossa tumors, particularly in radiated survivors of medulloblastoma.[48] In metabolic and degenerative disorders, the ataxia is usually progressive. Treatment is directed towards the underlying disorder, other associated features, and complications, particularly spasticity. These children require a multidisciplinary team approach with the involvement of several specialties including pediatrics, neurology, genetics, orthopedics, physiotherapy, and occupational therapy. Lioresal or diazepam may relieve spasticity, improve motility of the limbs, and combat pain. Specific treatments to counteract the offending metabolite, replace the dysfunctional enzyme, or vitamin therapy are needed in metabolic encephalopathies. Counseling the families and educating the public on these potentially preventable genetic disorders are very important in the management of these children.

In conclusion, unsteadiness and ataxia are common presentations of many acute and chronic disorders. Recognizing benign and non-neurological causes of unsteady gait is essential to avoid unnecessary investigations and hospital admission. Accurate assessment includes detailed history, examination, and then formulation of a differential diagnosis list to guide laboratory investigations. Cerebellar ataxia can be acute, chronic, progressive, or episodic. Acute cerebellar ataxia is the most common cause of childhood ataxia and usually results from drug ingestion or postinfectious cerebellar demyelination. Infections, brain tumors, and inherited disorders are important causes of progressive cerebellar ataxia. Intermittent ataxia should raise the suspicion of an underlying inborn error of metabolism. Acute exacerbations develop after high protein ingestion, concurrent febrile illness, or other physical stress. Sensory ataxia is less common in children and results from loss of posterior column sensory functions (proprioception and vibration sense). Common causes of acute cerebellar ataxia are mostly reversible. Treatment of chronic and progressive disorders depends on the underlying etiology. Counseling the families and educating the public of potentially preventable genetic disorders are very important in our region.

References

1. Jan MM. Perception of pediatric neurology among non-neurologists. *J Child Neurol* 2004; 19: 1-5.
2. Jan MM, Hasanain FH, Al-Dabbagh AA. Infant and child safety practices of parents. *Saudi Med J* 2000; 21: 1142-1146.
3. Jan MM. Clinical review of pediatric epilepsy. *Neurosciences* 2005; 10: 255-264.
4. Jan MM. Pediatric neuro-developmental and behavioral disorders: practitioner's perspectives. *Neurosciences* 2005; 10: 149-154.
5. Jan MM. The hypotonic infant: clinical approach. *J Pediatr Neurol* 2007; 5: 181-187.
6. Jan MM, Al-Buhairi AR, Baeesa SS. Concise outline of the nervous system examination for the generalist. *Neurosciences* 2001; 6: 16-22.
7. Jan MM, Khalifa MA. Nervous System and Development. In: Alhowasi MN, editors. Manual of clinical pediatrics (for medical students & postgraduate doctors). 3rd ed. Riyadh, Saudi Arabia: Medical Book House; 2004. p. 130-76.

8. Jan MM, Fida NM. Attitudes of medical students toward pediatric neurology. *Ped Neurol* 2002; 27: 106-110.

9. Al-Asnag MA, Jan MM. Influence of the clinical rotation on intern attitudes toward pediatrics. *Clin Pediatr (Phila)* 2002; 41: 509-514.

10. Jan MM, Wazzan OM, Baeesa SS. Impressions and experiences of non-neurologists in neurology. *Neurosciences* 2005; 10: 272-276.

11. Jan MM. Approach to Children with suspected neurodegenerative disorders. *Neurosciences* 2002; 7: 2-6.

12. Ryan MM, Engle EC. Acute ataxia in childhood. *J Child Neurol* 2003; 18: 309-316.

13. Jan MM. Neurological examination of difficult and poorly cooperative children. *J Child Neurol* 2007; 22: 1209-1213.

14. Gieron-Korthals MA, Westberry KR, Emmanuel PJ. Acute childhood ataxia: 10-year experience. *J Child Neurol* 1994; 9: 381-384.

15. Addas BM, Jan MM. Cerebellar abscess due to Listeria monocytogenes. *Saudi Med J* 2002; 23: 226-228.

16. Liu GT, Urion DK. Pre-eruptive varicella encephalitis and cerebellar ataxia. *Pediatr Neurol* 1992; 8: 69-70.

17. Connolly AM, Dodson WE, Prensky AL, Rust RS. Course and outcome of acute cerebellar ataxia. *Ann Neurol* 1994; 35: 673-679.

18. Rust RS. Multiple sclerosis, acute disseminated encephalomyelitis, and related conditions. *Semin Pediatr Neurol* 2000; 7: 66-90.

19. Jan MM. An unusual cause of isolated trochlear nerve palsy. *Neurosciences* 2007; 12: 149-151.

20. Jan MMS. Childhood multiple sclerosis. *J Pediatr Neurol* 2005; 3: 131-136.

21. Echenne B, Gras M, Astruc J. Vertebro-basilar arterial occlusion in childhood-Report of a case and review of the literature. *Brain Dev* 1983; 5: 577-581.

22. Schoenberg BS, Mellinger JF, Schoenberg DG. Cerebrovascular disease in infants and children: a study of incidence, clinical features, and survival. *Neurology* 1978; 28: 763-768.

23. Boltshauser E, Deonna T, Hirt HR. Myoclonic encephalopathy of infants or "dancing eyes syndrome." Report of 7 cases with longterm follow-up and review of the literature (cases with and without neuroblastoma). *Helv Paediatr Acta* 1979; 34: 119-133.

24. Case records of the Massachusetts General Hospital. Weekly clinicopathological exercises. Case 27-1995. A 29-month-old girl with worsening ataxia, nystagmus, and subsequent opsoclonus and myoclonus. *N Engl J Med* 1995; 333: 579-586.

25. Jan MM. Cerebral palsy: comprehensive review and update. *Ann Saudi Med* 2006; 26: 123-132.

26. Pollack IF. Brain tumors in children. *N Engl J Med* 1994; 331: 1500-1507.

27. Jan MM. Updated overview of pediatric headache and migraine. *Saudi Med J* 2007; 28: 1324-1329.

28. Freund HJ. Differential effects of cortical lesions in humans. *Ciba Found Symp* 1987; 132: 269-281.

29. Moin M, Aghamohammadi A, Kouhi A, Tavassoli S, Rezaei N, Ghaffari SR, et al. Ataxia-telengiectasia in Iran: clinical and laboratory features of 104 patients. *Pediatr Neurol* 2007; 37: 21-28.

30. Koht J, Tallaksen CM. Cerebellar ataxia in the eastern and southern parts of Norway. *Acta Neurol Scand* 2007; 187: 76-79.

31. Coman DJ, Sinclair KG, Burke CJ, Appleton DB, Pelekanos JT, O'Neil CM. Seizures, ataxia, developmental delay and the general paediatrician: glucose transporter 1 deficiency syndrome. *J Paediatr Child Health* 2006; 42: 263-267.

32. Bahassan NA, Jan MM. Ketogenic diet: update and application. *Neurosciences* 2006; 11: 235-240.

33. Brenner M, Oakley C, Lewis DW. Unusual headache syndromes in children. *Curr Pain Headache Rep* 2007; 11: 383-389.

34. Delanoe C, Sebire G, Landrieu P, Huault G, Metral S. Acute inflammatory demyelinating polyradiculopathy in children: clinical and electrodiagnostic studies. *Ann Neurol* 1998; 44: 350-356.

35. Cerisola-Cardoso A, Capote-Moreira G, Scavone-Mauro C. [Guillain-Barre syndrome in pediatrics. Different forms of presentation and difficulties involved in reaching an early diagnosis] *Rev Neurol* 2007; 44: 725-732. Spanish.

36. Mori M, Kuwabara S, Fukutake T, Yuki N, Hattori T. Clinical features and prognosis of Miller Fisher syndrome. *Neurology* 2001; 56: 1104-1106.

37. Huang CC, Liu CC, Chang YC, Chen CY, Wang ST, Yeh TF. Neurologic complications in children with enterovirus 71 infection. *N Engl J Med* 1999; 341: 936-942.

38. Hayward K, Jeremy RJ, Jenkins S, Barkovich AJ, Gultekin SH, Kramer J, et al. Long-term neurobehavioral outcomes in children with neuroblastoma and opsoclonusmyoclonus-ataxia syndrome: relationship to MRI findings and anti-neuronal antibodies. *J Pediatr* 2001; 139: 552-559.

39. Laug WE, Siegel SE, Shaw KN, Landing B, Baptista J, Gutenstein M. Initial urinary catecholamine metabolite concentrations and prognosis in neuroblastoma. *Pediatrics* 1978; 62: 77-83.

40. Weiss S, Carter S. Course and prognosis of acute cerebellar ataxia in children. *Neurology* 1959; 9: 711-721.

41. Tenembaum S, Chamoles N, Fejerman N. Acute disseminated encephalomyelitis. A long-term follow-up study of 84 pediatric patients. *Neurology* 2002; 59: 1224-1231.

42. Hynson JL, Kornberg AJ, Coleman LT, Shield L, Harvey AS, Kean MJ. Clinical and neuroradiologic features of acute disseminated encephalomyelitis in children. *Neurology* 2001; 56: 1308-1312.

43. North K, de Silva L, Procopis P. Brainstem encephalitis caused by Epstein-Barr virus. *J Child Neurol* 1993; 8: 40-42.

44. Mitchell WG, Snodgrass SR. Opsoclonus-ataxia due to childhood neural crest tumors: a chronic neurologic syndrome. *J Child Neurol* 1990; 5: 153-158.

45. Asbury AK. New concepts of Guillain-Barré syndrome. *J Child Neurol* 2000; 15: 183-191.

46. Bradshaw DY, Jones HR Jr. Guillain-Barré syndrome in children: clinical course, electrodiagnosis, and prognosis. *Muscle Nerve* 1992; 15: 500-506.

47. Beckung E, Carlsson G, Carlsdotter S, Uvebrant P. The natural history of gross motor development in children with cerebral palsy aged 1 to 15 years. *Dev Med Child Neurol* 2007; 49: 751-756.

48. Huber JF, Bradley K, Spiegler B, Dennis M. Long-term neuromotor speech deficits in survivors of childhood posterior fossa tumors: effects of tumor type, radiation, age at diagnosis, and survival years. *J Child Neurol* 2007; 22: 848-854.