

Evaluating the Child with Unsteady Gait

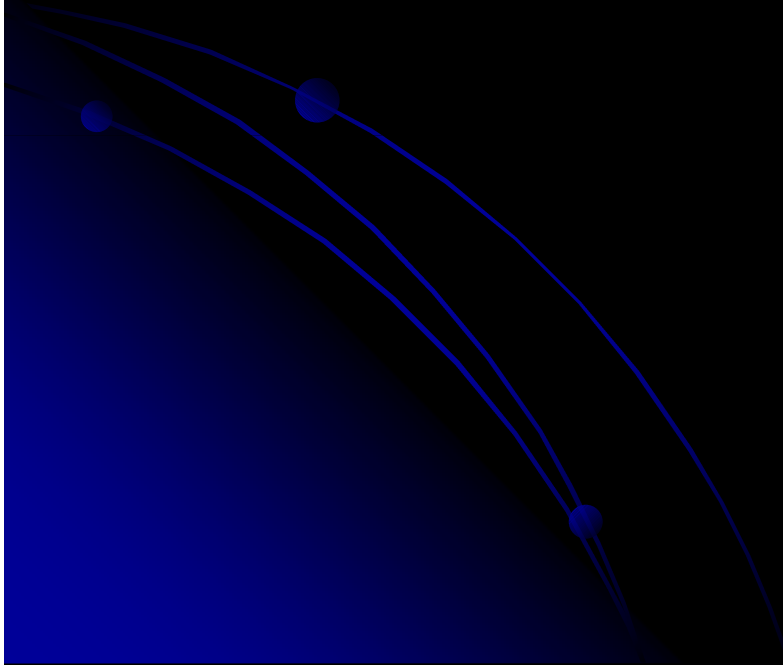
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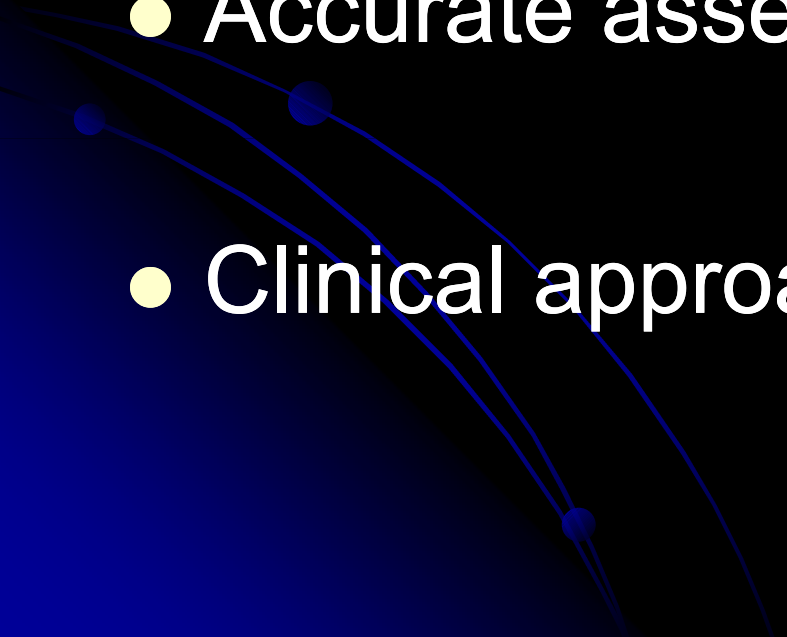
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لا حول و لا قوة إلا بالله! اللهم هون عليهم و كن معهم!
اللهم أطف بهم! اللهم لا تؤاخذنا على غفلتنا و قسوة قلوبنا!
يا أكرم الأكرمين! حسبنا الله ونعم الوكيل



ABNORMAL GAIT

- Common presentation of a variety of acute and chronic disorders
 - Detailed history and exam
 - Accurate assessment
 - Clinical approach to the unsteady child
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UNSTEADY GAIT

- Not always neurological
 - Simple injuries and musculoskeletal
 - Recognize benign causes
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Causes of unsteady gait in children

Foot deformity

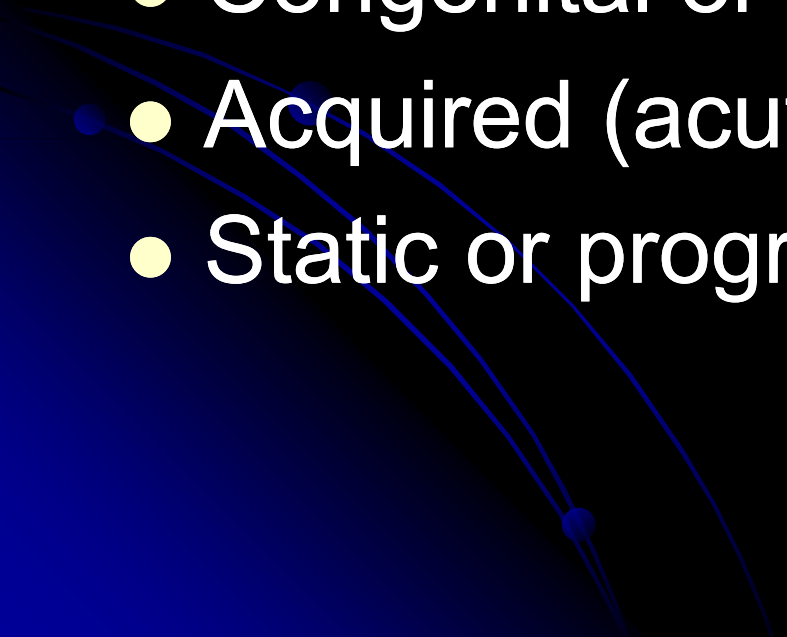
Skeletal abnormalities (ankle, knee, or hip joint)

Antalgic gait (due to pain)

Migraine (basilar, benign paroxysmal vertigo)

Raised intracranial pressure (hydrocephalus)

ATAXIA

- Lacking order in Greek
 - Cerebellar, motor, sensory
 - Congenital or acquired
 - Acquired (acute, chronic, episodic)
 - Static or progressive
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Causes of ataxia in children

Cerebellar Ataxia

Congenital / Genetic

Traumatic (contusion, hemorrhage, concussion, VB dissection)

Toxic and Drugs (alcohol, antihistamines, anticonvulsants)

Infectious/immune-mediated (chicken pox, ADEM, MS)

Malignancy (medulloblastoma, neuroblastoma)

Paraneoplastic (opsoclonus-myoclonus syndrome)

Vascular (stroke, hypertension, AV malformation, bleeding)

Degenerative (ataxia telangiectasia)

Post-ictal (epileptic ataxia)

Causes of ataxia in children

Sensory Ataxia

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

Other

Paretic ataxia (weakness due to U or LMNL)

Functional ataxia (munchausen by proxy)

CLINICAL EVALUATION

History

- Refusal to walk or wide-based or drunken gait
- Tremor, head titubation, trunk, dysarthria
- Exclude serious causes (infections and tumors)
- Symptoms of infection, trauma, exposure to drugs or household chemicals
- Headache, vomiting, diplopia

CLINICAL EVALUATION

History

- Acute, chronic, progressive ataxia
- Intermittent ataxia
- Precipitated by infections or drugs (VPA)
- Recent immunizations
- Past medical and family histories

Types of cerebellar ataxia in childhood

Acute

Trauma

Toxic and Drugs

Seizure related (post-ictal, nonconvulsive status)

Infections / Postinfectious

Vascular (stroke, hypertension, AV malformation, bleeding)

Malignancy (medulloblastoma, neuroblastoma)

Paraneoplastic (Opsoclonus-myoclonus syndrome)

Functional

Types of cerebellar ataxia in childhood

Chronic

Congenital (hypoplasia, Dandy-Walker, Chiari)

Post-traumatic

Post-meningitis / encephalitis

Post-tumor resection or radiation

Hypoxic-ischemic insult

Types of cerebellar ataxia in childhood

Progressive

Friedreich ataxia

Ataxia Telangiectasia

Sphingolipidosis (gangliosidosis, Niemann-Pick disease)

Leukodystrophies (PM, Krabbe, metachromatic)

Mitochondrial disorders (Leigh disease, MERF)

Neuronal ceroid-lipofuscinosis

Progressive myoclonic epilepsies (Lafora, UL disease)

Congenital defect of glycosylation

Abetalipoproteinemia

Types of cerebellar ataxia in childhood

Recurrent / Intermittent

Migraine (basilar, benign paroxysmal vertigo)

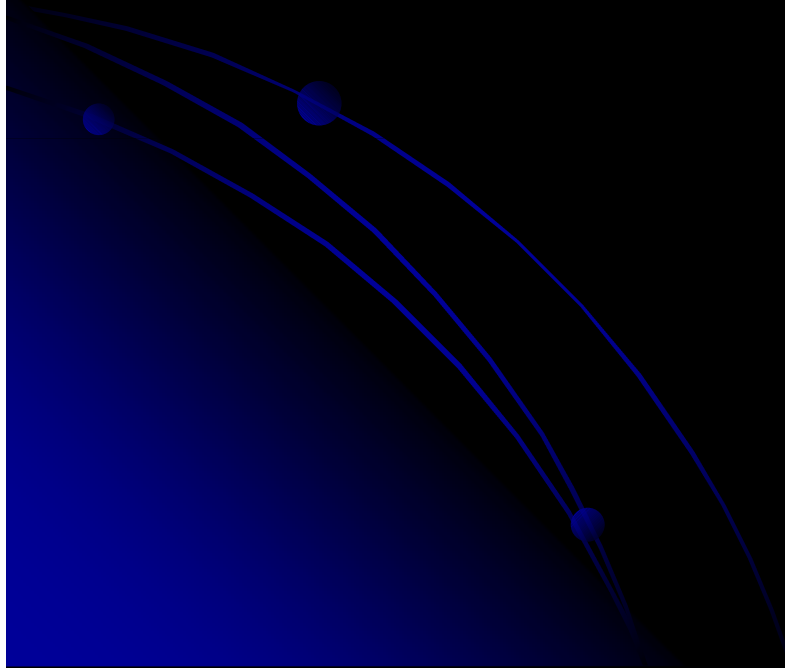
Genetic (autosomal dominant episodic ataxias)

Metabolic (AA, urea cycle, mitochondrial)

CLINICAL EVALUATION

Examination

- Often uncooperative and irritable
- Observe for important signs
- Trauma, meningeal signs
- Eyes (pupils, telangiectasia, nystagmus, papilledema)
- Cranial nerves (tumors or hydrocephalus)



CLINICAL EVALUATION

Examination

- Altered responsiveness (drug ingestion, toxic exposure)
- Irritability (meningitis, encephalitis, OM syndrome)
- Motor exam (weakness, hypotonia, incoordination)
- Gait (off balance with eyes open and worse with closure)
- Walking on a straight line (sway towards the side)
- Tandem walk is more sensitive
- Dysmetria, poor coordination with under or overshooting

CLINICAL EVALUATION

Examination

- Dysdiadochokinesia
- Finger nose, heel shin, foot tap
- Intention tremor (increases at target)
- Pendular deep tendon reflexes
- Sensory exam (vibration and position)
- Romberg sign (with eye closure)

CEREBELLAR ATAXIA

Acute cerebellar ataxia

- Postinfectious is most common (40%)
- Antecedent illness 1-3 weeks (varicella)
- Symptoms are maximal at onset (truncal)
- ADEM (post viral illness or vaccination)
- Systemic features, seizures, CNS deficits
- MS (optic neuritis and long-tract signs)

CEREBELLAR ATAXIA

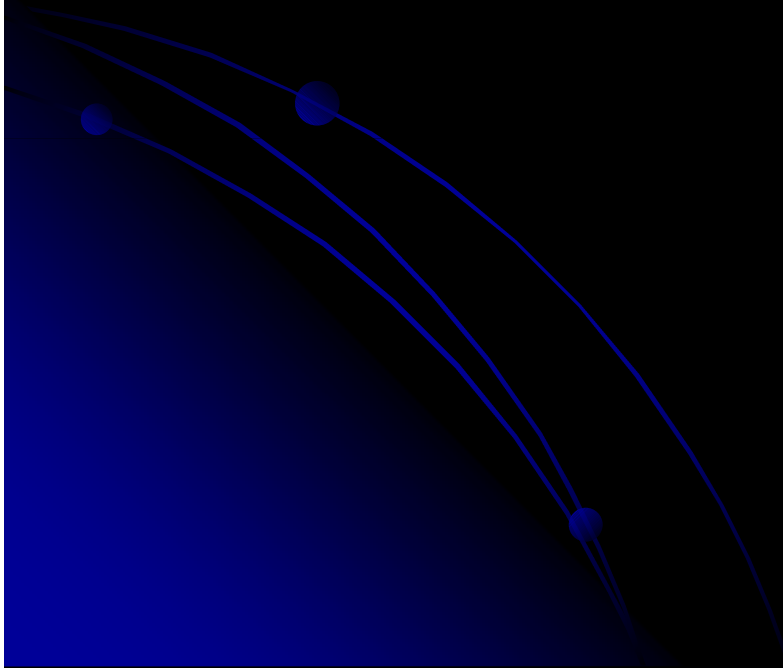
Acute cerebellar ataxia

- Head injuries (concussion, contusion, bleed)
- Vascular insult
- Opsoclonus-myoclonus syndrome
- Postinfectious or occult neuroblastoma
- Drugs (AEDs, benzos, alcohol, antihistamines)
- Lethargy, confusion, inappropriate behavior

Chronic cerebellar ataxia

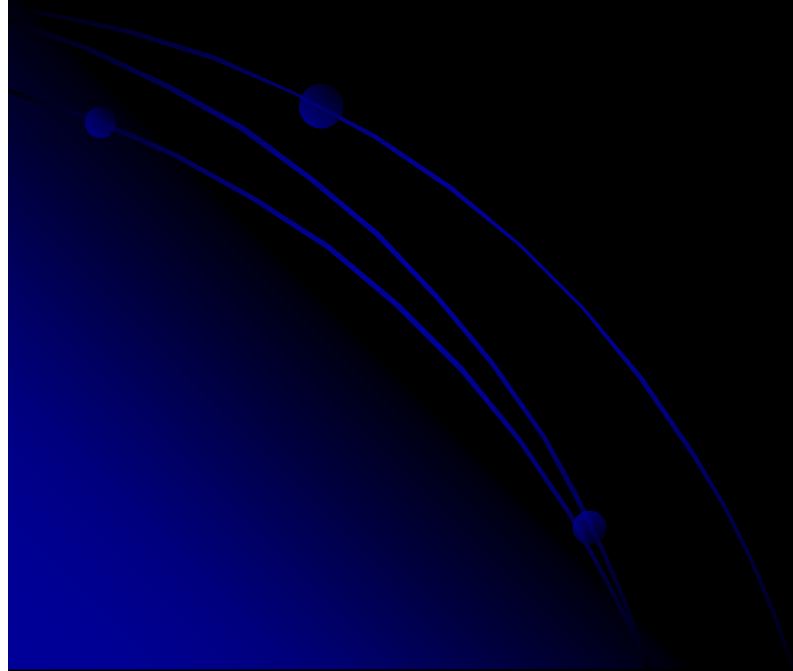
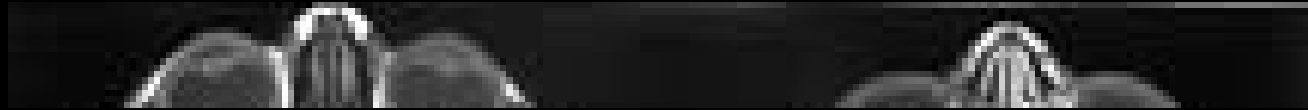
- Static (non-progressive) anomalies or insults
- Congenital (hypoplasia, vermal aplasia, chiari)
- Insults (post trauma, infection, hypoxia, ischemia)
- Motor, developmental and cognitive deficits
- Improvements (severity, other deficits, therapy, rehabilitation)

Cerebellar Hypoplasia



Progressive cerebellar ataxia

- Tumors (50% arise from brain stem, cerebellum)
- Progressive ataxia, features of increased ICP, personality change
- Frontal lobes (frontocerebellar associative fibers)
- Metabolic and degenerative disorders
 - Ataxia telangiectasia, spinocerebellar ataxias (Friedreich)
 - Glucose transporter deficiency syndrome



Recurrent cerebellar ataxia

- Migraine and periodic syndromes
- Vomiting, ataxia, vertigo (\pm headache)
- AD episodic ataxias (mutations in ion channel)

- Inborn error of metabolism (with stupor)
- Acute exacerbation (high protein meal, febrile illness, drugs, physical stress)
- Rare presentation (late infantile and juvenile partial forms)

Causes of intermittent ataxia

Migraine and migraine variants

Basilar migraine

Benign paroxysmal vertigo

Benign paroxysmal torticollis of infancy

Alternating hemiplegia of childhood

Causes of intermittent ataxia

Genetic disorders

Episodic ataxia type 1 (with myokymia)

Episodic ataxia type 2 (acetazolamide responsive)

Episodic ataxia types 3 and 4

Episodic ataxia with paroxysmal dystonia

Causes of intermittent ataxia

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

SENSORY ATAXIA

- Wide-based, “steppage” gait
 - Worsen with the eyes closed
 - Posterior column, roots, or peripheral nerves
 - Loss of proprioception and vibration sense
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- Romberg’s sign and decreased tendon reflexes
 - GBS (Miller Fisher variant)
 - Ataxia, areflexia, and ophthalmoplegia

INVESTIGATIONS

- Exclude serious conditions
- History and exam (screening tests)
- Drug screen
- CT (posterior fossa artifacts)
- MRI (ADEM, MS, encephalitis)

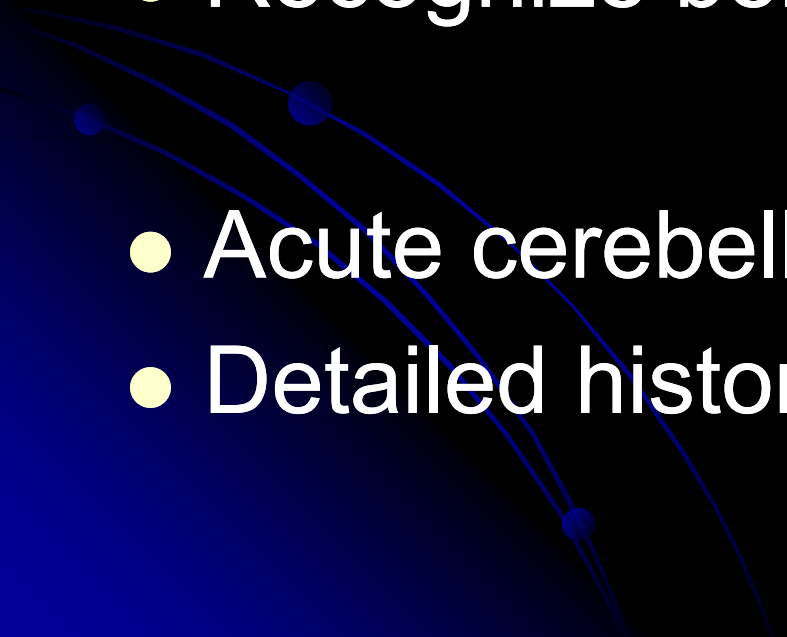
INVESTIGATIONS

- CSF (encephalitis, GBS, MS)
- EMG (GBS, sensory ataxia)
- Urinary catecholamine metabolites, CT chest and abdomen
- Serum IgA, IgE, and alpha fetoprotein
- Metabolic workup
- DNA tests and enzyme assays

MANAGEMENT

- Reversible conditions
- ADEM, MS (corticosteroids)
- GBS (IVIg, plasmapheresis)
- Ingestions (antidote, chelation, dialysis)
- Metabolic (specific treatments to counteract the offending metabolite, replace the dysfunctional enzyme, or vitamin therapy)
- Genetic counseling

CONCLUSIONS

- Common presentation of many acute and chronic disorders
 - Recognize benign and non-CNS causes
 - Acute cerebellar ataxia is most common
 - Detailed history and exam to guide tests
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Review Article

