Acute Ataxia

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EPNS Training Course March 2015 Budapest

„Acute"

- Not defined by consensus
- Ryan and Engle (2003) evolution time < 72 hours
- **Symptoms within hours („over night"), < 2 days**

Ataxia

- „Lack of order"
- Ataxia ≠ cerebellar disorder
- Efferent pathways involved
- Afferent pathways involved → sensory ataxia(s)
  - chronic - acute
  - hereditary - acquired
- Vestibular
- Psychogenic

Sensory versus cerebellar ataxia

- No dysarthria
- «no» oculomotor deficits
- Romberg test abnormal
  eyes closed – no visual compensation – ataxia worse

Neuropediatrics 2013;44:127-141

Review Article

Acute Ataxia in Children: Approach to Clinical Presentation and Role of Additional Investigations

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Table 1 Causes of acute pediatric ataxia

Acute cerebellar ataxia	Infectious/immune mediated	Acute postinfectious cerebellar ataxia
		Acute cerebellitis
		Acute disseminated encephalomyelitis
		Multiple sclerosis
	Intoxications	
	Paraneoplastic	Opsoclonus-myoclonus syndrome
	Traumatic	Postconcussion ataxia
		Traumatic vertebral dissection
	Vascular	Ischemic stroke
		Hemorrhagic stroke
	First event intermittent ataxia*	Maple syrup urine disease
		Pyruvate dehydrogenase deficiency
		Urea cycle disorders
		Glucose transporter type 1 deficiency
	First episode episodic ataxia	
Acute vestibular ataxia	Migraine related	Vestibular migraine, first episode
		Benign paroxysmal vertigo
	Acute unilateral vestibular dysfunction	Vestibular neuritis
		Labyrinthitis
		Vestibular contusion
Acute sensory ataxia	Inflammatory	Guillain-Barré syndrome
		Miller Fisher syndrome
Acute epileptic pseudoataxia		
Acute psychogenic ataxia		

Ataxias in childhood - Categories

according presentation and course (~ arbitrary)

- Acute ataxia (< 2-3 days)
- Subacute ataxia

- Episodic ataxia
  [historic term, implies dominant channelpathy]

- Intermittierent ataxia

- Congenital non-progressive ataxia
- Chronic progressive ataxia

Distinction not always possible at onset

Repeated events of acute ataxia

Ad terminology:
„Episodic ataxia“ implies channelopathies
→ »intermittent»

DD

- Repeated intoxications («external»)
- Benign paroxysmal vertigo
- Basilar migraine
- Metabolic disorders («internal intoxication»)
  (usualy in catabolic situation)
- Episodic ataxias EA1, EA2..
- ……

Ataxia in metabolic disorders

- Mostly in catabolic situations (infection, fever..)
- «internal intoxication» (simplified)
  → usually ataxia *plus*
- **Typical examples**
  - urea cycle disorders
  - amino acid disorders (as MSUD, maple syrup urine disease)
  - organic acidurias
- **Investigations**
  - «routine» lab incl lactate, ammonia, blood gases
  - plasma: amino acids, acylcarnitine, homocysteine
  - urine: organic acids
- **MRI** – my be helpful (pattern recognition)

Acute postinf. cerebellar ataxia	**Acute cerebellitis**
No neuroimaging correlate	Neuroimaging correlate +
Isolited ataxia	Often additional symptoms ev. oedema - hydrocephalus herniation
General outcome favorable	Overall prognosis less favorable

Distinction justified ? arbitrary ?
Rather a spectrum – a continuum ?
In praxi distinction often helpful:
→ Different management

Examination Points to consider in acute ataxia

- Consciousness Responsiveness
  Behavior
- **Ataxia ? Pure ? Plus ?**
  Weakness ? (Pareses)
- Ataxia (Dys-metria)
  Trunk ? Limbs ? Tongue ? Ocular movements ?
- Focal findings – asymmetry ?
- Ocular movements
- Head impulse test
- Red flags ?

Acute ataxia - what is „common“ ?

Common
- Acute postinfectious cerebellar ataxia
- Intoxication
- ADEM

Rare
- Cerebellitis
- Opsoclonus - Myoclonus Syndrome
- Stroke
- Varia

NOT PRESENTING AS *ACUTE* ATAXIA
- Cerebellar tumor
- Meningitis

Intoxication

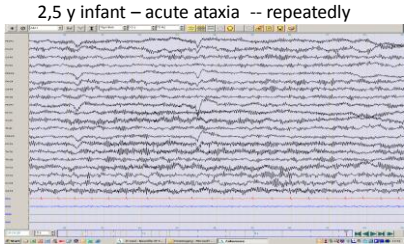
Age peaks
- Infancy
  also as Münchhausen by proxy
- [Teenage (alcohol, drugs, suicidal attempts…)]

Consider circumstances – awareness

Red flags ? Consciousness impaired

Investigations – consider
- „Tox" screening (serum, urine)
- EEG

2,5 y infant – acute ataxia -- repeatedly



Increased beta-activity due to benzodiazepin medication

[Münchhausen by proxy]

Acute (postinfectious) cerebellar ataxia

- Occurrence
  post viral and non-viral infections
  (varicella, EBV, mumps, parvovirus…)
  in ~20% no previous infection
- Age
  predominant in young children (~2-5 y)
  but at any age reported
- Onset
  acute, „over night", max.symptoms in 1-2 days
- Course
  spontaneous improvement over days to few weeks
  no relapses (rare exceptions to the rule)

Acute cerebellar ataxia

- Symptoms
  Ataxia trunk > limbs
  Nystagmus, dysarthria : not consistant
  RED FLAGS: papilledema, vomiting, strabismus
  impaired level of consciousness
- Additional investigations
  EEG, CSF, imaging: usually normal (→ no strict indication)
  ((Serology ?))
- Course: remission in days – weeks
- Recovery: favorable (exceptions)
- Treatment : no steroids - wait and see

RED FLAGS

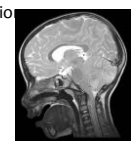
…what is beyond pure ataxia

Cerebellitis

Variable signs and symptoms – spectrum

Focal – Hemi- -- entire cerebellum

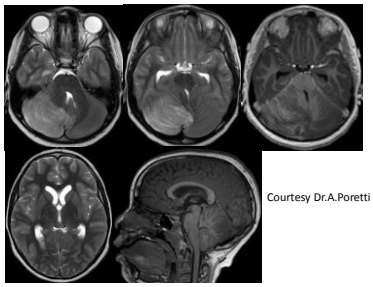
Swelling (oedema)
→ compression brain stem / herniation
→ acute obstructive hydrocephalus
(→ NO Lumbart Puncture
(→ surgical decompression



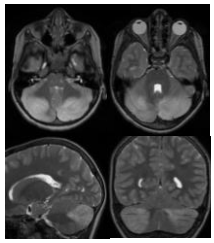
Extreme end of spectrums
Near fatal cerebellar swelling

Fulminant cerebellitis: a fatal clinically isolated syndrome. Kamate 2009
Near-fatal cerebellar swelling caused by…Burri 2003
Acute fatal parainfectious cerebellar swelling…Roulet Perez 1993

Hemi-Cerebellitis – midline shift - hydrocephalus - Mycoplasma infection



Courtesy Dr.A.Poretti



Cerebellitis bilateral

Grey and white matter
No significant swelling

6 year old
History of non-specific findings, vomiting, fever,
Reduced condition

Increasing headache
Ataxia ? (not examined!)

Spontaneous recovery

Opsoclonus – Myoclonus Syndrome

- Very rare....but diagnosis important for treatment
- Characteristic symptoms
- Diagnosis can be made by history / clinical examination

- Synonyma
  Kinsbourne syndrome [1962, 6 infants, collected P. Sandifer]
  Myoclonic encephalopathy of infancy
  Dancing eyes syndrome

- Age predilection
  Months to 3 years

Opsoclonus - Myoclonus Syndrome

Pathogenesis

- „postinfectious"…no obvious other cause
- „paraneoplastic" associated with neuroblastoma or ganglioneuroma

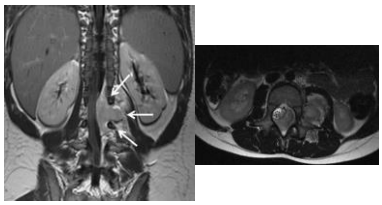
Additional investigations

- Search for neuroblastoma

- Characteristic symptoms
  - Ataxia (usually no longer able to sit)
  - Myoclonia (easy to feel)
  - Opsoclonus (inconsistant)
  - Irritability, sleep disturbances
- **„Atypical" presentations (~20%)**
- Pathogenesis
- Course (untreated)
  Usually prolongued…over weeks and months
  Relapses with infection
  Majority of patients (untreated) with residual problems !!
- Investigations (CSF,EEG,MRI normal at onset) →clinical dg
- Treatment... („immunosuppressive")
- Long-term course ...
  European protocoll for diagnosis und treatment

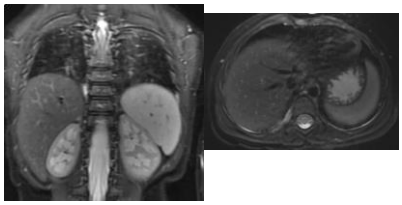
Infant with OMS

Ganglioneuroma paravertebral – intraspinal extension
no neurological deficit

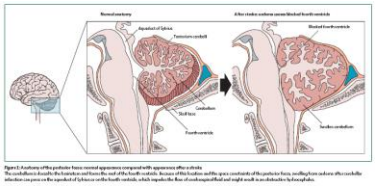


Subdiaphragmatic neuroblastoma in atypical OMS

(Atypical – no opsoklonus, protracted onset)
Excelllent response to immunosuppressive tp, not operated



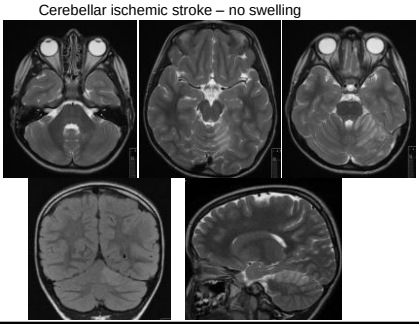
Cerebellar swelling with potential brain stem compression and herniation, possible in
- stroke
- cerebellitis



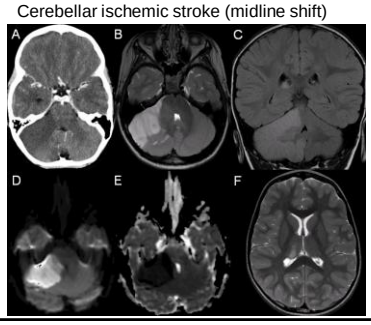
Diagnosis and initial management of cerebellar infarction

Lancet Neurology 2008;7:951-64

Cerebellar ischemic stroke – no swelling



Cerebellar ischemic stroke (midline shift)



Additional investigations

- Individual work-up targeted on the basis of differentiated clinical suspicion

- Acute isolated ataxia without red flags
- Acute ataxia with red flags
  high value of neuroimaging
  Consider ev. risk of LP (cerebellitis, stroke)
  ? CSF, serological tests etc
  further investigations depending on MRI, course…
- ? Intoxication: EEG, body fluid collection
- ? Metabolic disorder – collect body fluids in acute stage
- OMS
  Investigations according protocol
- Functional (psychogenic) disorder – no further tests !!
- ….

Acute disseminated encephalomyelitis ADEM

- Age at onset median 5-8 years
- Acute (subacute) multifocal inflammatory demyelinating process
- Commonly preceded by an infection (usually viral)
- Headache, fever
- Transitory and self-limiting
- Accompanied by encephalopathy
  (according new consensus)

Definition ADEM (monophasic)

- First event
- Acute or subacute onset
- Clinical presentation: polyfocal symptoms including «encephalopathy»,
  defined as:
  behavioral change, e.g. confusion, irritability..
  Alteration of consciousness, e.g. lethargy, coma
- Event followed by improvement
- New or fluctuationg signs and symptoms within 3 months considered part of acute event
- Neuroimaging…

ADEM

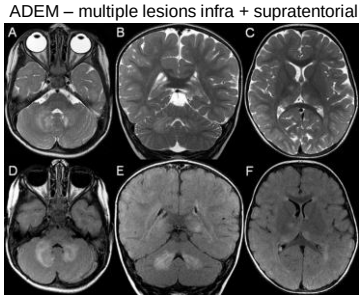
- **Imaging**
  bilateral mulifocal subcortical white matter lesions
  cortex not involved
  central grey matter nuclei (bg,thalami) often affected
  spinal cord: lesions common, often extensive
- **CSF**
  Protein mostly elevated (mild to moderate)
  Pleocytosis common (lymphocytic)
  Oligoclonal bands usually not present (0-30%)
- **Treatment**
  → steroids

Clinical and neuroradiological differences of paediatric acute disseminating encephalomyelitis with and without antibodies to the myelin oligodendrocyte glycoprotein

Baumann et al JNNP 2015

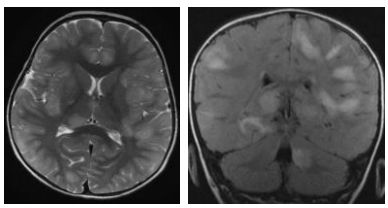
Children with ADEM and MOG antibodies
- Better outcome
- MRI brain - large, bilateral, widespread lesions
- MRI spine - often extensive longitudinal lesions

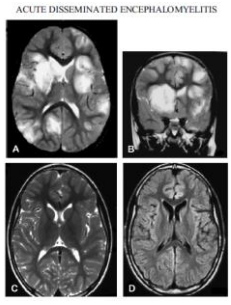
ADEM – multiple lesions infra + supratentorial



Multifocal lesions: supra- and infratentorial
central grey matter + subcortical white matter

ADEM T2w FLAIR





ADEM in 3 year old following respirat inf.

A,B – acute
C, D – follow-up

From
Tennenbaum 2013

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