Acute Ataxia

Eugen Boltshauser
Emeritus – Department of Pediatric Neurology
Children’s Hospital Zürich
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

Table 1: Causes of neuropediatric ataxia

<table>
<thead>
<tr>
<th>Scale vestibular ataxia</th>
<th>Sensory/projection cerebellar ataxia</th>
<th>Ataxia spinocerebellar</th>
<th>Acute hemispheric syndromes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infarction</td>
<td>Hydrocephalus</td>
<td>Brainstem damage</td>
<td>Stroke</td>
</tr>
<tr>
<td>Traumatic</td>
<td>Spinal cord injury</td>
<td>Tumors</td>
<td>Meningitis</td>
</tr>
<tr>
<td>Tumors, metastases</td>
<td>Multiple sclerosis</td>
<td>Multiple sclerosis</td>
<td>Multiple sclerosis</td>
</tr>
<tr>
<td>Acute encephalopathy</td>
<td>Multiple sclerosis</td>
<td>Multiple sclerosis</td>
<td>Multiple sclerosis</td>
</tr>
</tbody>
</table>

Neuropediatrics 2013;44:127-141

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

Andrea Piovesi, J.J. and E. Benson, T. Heyer, A. G. M. Hilman and G. Böckenhauer
Ataxias in childhood - Categories
according presentation and course (~ arbitrary)

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

• Episodic ataxia
  [Historic term, implies dominant channelopathy]
• Intermittent 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”) (usually 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 (as MSUD, maple syrup urine disease)
  - Amino acid disorders (as organic acidurias)

• Investigations
  - “routine” lab incl lactate, ammonia, blood gases
  - Plasma: amino acids, acylcarnitine, homocysteine
  - Urine: organic acids

• MRI – may be helpful (pattern recognition)
Acute postinf. cerebellar ataxia

- Acute cerebellitis
- No neuroimaging correlate
- Isolated ataxia
- General outcome favorable

- Neuroimaging correlate +
- Often additional symptoms ev. edema - hydrocephalus herniation
- 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 consistent
  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 (edema)
  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... Roquet Perez 1993
Hemi-Cerebellitis – midline shift - hydrocephalus - Mycoplasma infection

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 (inconsistent)
- Irritability, sleep disturbances

* “Atypical” presentations (~20%)

- Pathogenesis
  - Course (untreated)
    - Usually prolonged...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 protocol for diagnosis und treatment

Infant with OMS

Ganglioneuroma paravertebral – intraspinal extension
no neurological deficit
Subdiaphragmatic neuroblastoma in atypical OMS

(Atypical – no opsklonus, protracted onset)
Excellent response to immunosuppressive tp, not operated

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

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 fluctuating signs and symptoms within 3 months considered part of acute event
- Neuroimaging...

ADEM

- Imaging
  - bilateral multifocal 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 JNPP 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 in 3 year old
following respirat inf.
A,B – acute
C, D – follow-up

From
Tennenbaum 2013
References ad ADEM

- Krupp LB, Rennell B, Tenembaum S
  Consensus definitions proposed for pediatric multiple sclerosis and related conditions. Neurology 2007;68:57-12
- Tenembaum S et al
  Acute disseminated encephalomyelitis. Neurology 2007;68:S7-S12
- Parrish JB, Yeh EA
  Acute disseminated encephalomyelitis. Rev Neurol 2012;72:1-26
- Pohl D, Tenembaum S
  Treatment of acute disseminated encephalomyelitis. Curr Treat Options Neurol 2012;14:264-275
- Tennenbaum S
- Alper G

Additional references

- Poretti et al
  Acute ataxia in children. Approach to clinical presentation and role... Neuropediatrics 2013;44:127-41
- Desai J and Mitchell WG
  Acute cerebellar ataxia, acute cerebellitis, and opsoclonus-myoclonus syndrome
- Hero B, Schlieker B
  Update on pediatric opsoclonus-myoclonus syndrome. Neuropediatrics 2013;44:324-29