Acute cranial nerve deficits

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EPNS Training Course March 2015 Budapest

Inclusion - Exclusion

• Pediatric focus
• Acute < 2-3 days
• Not considered – acute visual loss, acute hearing loss
• «Acute» - sometimes a longstanding problem is only recently realised (Pseudo-acute)
Acute ➔ «categories»?

- Trauma
- Infection – inflammation
- Haemorrhage
- Intracranial pressure
- Tumor
- Toxic

Acute oculomotor nerve palsy
Acute IIIrd nerve palsy

«Ophthalmoplegic migraine»
(re-classified as cranial neuralgia)

• Observed in (young) children
• Onset usually associated with transient headache, nausea, vomiting
• III rd nerve palsy, pupil mostly not spared (variable)
• Outcome – spontaneous recovery in a few weeks
• Treatment – wait and see
• Recurrences possible
• MRI – enhancement of thickened intracisternal portion of oculomotor nerve

Enhancement of cisternal portion of oculomotor nerve
11 year old girl
Acute IIIrd nerve palsy

IIIrd nerve thickened
Enhancement +

Painful Ophthalmoplegia – Tolosa Hunt Syndrome -
Cavernous sinus lesion

• Not an aetiological entity
• Initial cranial nerve dysfunction variable (isolated, combined)
  mostly III > VI … combinations
• MRI – high priority

Tolosa-Hunt syndrome (Painful ophthalmoplegia)
The International Headache Society introduced five criteria for the diagnosis of THS:
1. One or more episodes of unilateral orbital pain persisting for weeks if untreated
2. Paresis of one or more of the third, fourth, and/or sixth cranial nerves and/or demonstration of granuloma by MRI or biopsy
3. Paresis coincides with the onset of pain or follows it within 2 weeks
4. Pain and paresis resolve within 72 hours when treated adequately with corticosteroids
5. Other causes have been excluded by appropriate investigations

International Headache Society classification – open access 3rd ed – Cephalagia 2013;33(9)
Acute (isolated) mydriasis

- Isolated (no ptosis, normal ocular motility)
- Subjective complaint – feeling of glare (no pupillary contraction)
- Objective observation – parents, peers...

Causes
- Local drug (drops, patches...)
- Tonic pupil syndrome (Adie)
  - Unilateral > bilateral
  - Relative mydriasis in bright illumination
  - Poor to absent light reaction
  - Defective accommodation
  - Slow contraction to prolonged near-effort
  - Pupil constricts with pilocarpine (0.125%)

Benign condition – often spontaneous recovery
Acute abducens nerve palsy

Distinguish
Abducens nerve palsy Abduction deficit

Non-localizing
Parainfectious, trauma, diabetes, Increased intracranial pressure incl. Pseudotumor cerebri

Localizing
Pons – CP angle – Clivus – middle fossa – cavernous sinus, sup orbital fissure

Acute abducens palsy and pontine glioma

• Usually NOT isolated
  + other cranial nerve deficits and longtract signs

Cerebellar low grade glioma – obstructing fourth ventricle → hydrocephalus
Intrinsic pontine glioma – fourth ventricle displaced, but patent
→ no hydrocephalus

Acute benign abducens nerve palsy in infants

- Age group - usually infants
- «Idiopathic», «postinfectious», («post vaccination»)
- Isolated (» association with pontine glioma)
- Spontaneous recovery in weeks
- Recurrences possible

- Investigations – (a matter of «temperament»)
Acute facial nerve palsy

= „peripheral“ paresis (lower motorneuron affected)
In praxi: unilateral isolated
Anglo-american term
Bell's palsy = acute peripheral facial paresis of unknown origin

Red flags → further investigations
Paresis not isolated – subacute onset – bilateral
– central – Age < 2 years

Facial nerve paresis

Central
Forehead spared

Peripheral
All muscles affected

Right facial paresis
Hypoplasia M. depressor anguli oris right
### Acute peripheral facial palsy

#### Occurrence
- "Idiopathic" (Bell's palsy) ~ 50-70%
- Infectious
  - Neuroborreliosis (saisonal - Europe) ~ up to 30%
  - Viral (herpes..) ~ 10-20% (?)
- Skull base (petrous bone) - process
- Tumor intracranial (brainstem, cerebello-pontine angle)
- Hypertension (blood pressure)
- Leukaemia (very rare)
- Trauma
- Melkersson-Rosenthal Syndrome (OMIM %155900)
- Varia

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### Facial palsy

2nd «episode»

1st facial palsy 1 year ago

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### Melkersson-Rosenthal Syndrome

- Cheilitis
- Tongue changes not consistent (Lingua plicata)
Acute peripheral facial paresis

**HISTORY**
- Suggestion for ENT process?
- Tick bite? Erythema chron. migrans?
- Exposure to cold and wind?

**EXAMINATION**
- Focused neurological examination (isolated VII paresis?)
- Check ENT
- Blood pressure
- Optimal: grading of paresis (House Brackman) (or photo)

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**Facial nerve grading according to House, 1983, Laryngoscope**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Definition</th>
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<tbody>
<tr>
<td>I</td>
<td>Normal facial function in all areas.</td>
</tr>
<tr>
<td>II</td>
<td>Mild dysfunction: slight weakness noticeable only on close inspection. At rest, normal symmetry and tone. Motion some but normal movement of forehead, ability to close eye with minimal effort and slight asymmetry. No spasticity, contractions or hemicerebral spasm.</td>
</tr>
<tr>
<td>III</td>
<td>Moderate dysfunction: obvious but not disabling difference between involved and uninvolved areas. Movement slightly reduced or absent. Ability to close eyes with mild effort. Significant asymmetry. Patients with obvious translateral deviations, contractions, and hemicerebral spasms are Grade III regardless of the severity of facial weakness.</td>
</tr>
<tr>
<td>IV</td>
<td>Moderately severe dysfunction: obvious weakness and or disabling asymmetry. At rest; normal symmetry and tone. Motion an movement of forehead leading to obvious weakness with minimal effort. Symmetrical movement consists of slight to moderate weakness of mouth with minimal effort. Patients with eyelid, nose, and hemicerebral spasm receive grade IV regardless of degree of motor action.</td>
</tr>
<tr>
<td>V</td>
<td>Severe dysfunction: Only barely perceptible motion. At rest; prosthesis with drop of corner of mouth and occasional or absent nasolabial fold. Movement of forehead, in normal effort slight movement of buccal muscles of effort, slight movement of corner of mouth. Spasticity, contractions, and hemicerebral spasms usually absent.</td>
</tr>
<tr>
<td>VI</td>
<td>Total paresis: Less of tone, asymmetry, no motion, no spasticity, contracture, or hemicerebral spasm.</td>
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Memo ad Examination - Trigeminus nerve involved?

- Corneal reflex
  afferent – n. trigeminus
efferent – n. facialis bilateral

In marked paresis – look for innervation on healthy side!

ADDITIONAL EXAMINATION (IMAGING ?)
- not required in isolated unilateral VII paresis
- wait and see – further steps depend on course

TREATMENT (STEROIDS ?)
controversial issue - controversial publications
limitations: often small pediatric cohorts, retrospective, # grading steroids do not impair serological tests

„Steroids should be offered in new-onset Bell’s palsy” [< 72 hours]

Reconciling the clinical practice guidelines on Bell palsy from the AAO-HNSF and the AAN
Schwartz et al, Neurology 2014;82:1927-29
AAO-HNSF American Academy of Otolaryngology –Head and Neck Surgery Foundation
AAN recommendation ....oral steroids should be offered
AAO-HNSF recommendation ....clinicians should prescribe oral steroids
within 72 hours of onset of symptoms
**Rationale for steroid treatment**

Recovery in severe palsy often incomplete

Synkinesias when smiling
Smile – ptosis
Equal HBS grade 3

Inflammation → swelling of facial nerve at entry to internal auditory canal / facial canal → compression within bony structures

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**Practical management – Zürich children’s hospital**

**Additional investigations**
- always: blood film, serum sample for later serological tests
- if neuroborreliosis possible – discuss CSF examination
- no neuroimaging

**Treatment**
- steroids for 7 days (preferably within 48-72 h after onset)
- «ocularprotection» (eye drops, «artificial tears»)

**Information to child/parents**

Arrange short-term follow-up
Pediatric Neurology

MRI at presentation (age 2 months)

MRI 2 months later

Original article
Facial Nerve Paralysis in Children: Is It as Benign as Supposed?
Artane Biebl MD, Evelyn Lechner MD, Katarina Bruncek MD, Andrea Preisinger MD, Astrid Eisenkölbl MD, Klaus Schmidt MD, Dieter Forthner MD
Department of Pediatrics, Children's Hospital, Uni. Asch

Retrospective study, n=56

Pediatric Neurology 2014

Do Oral Steroids Aid Recovery in Children With Bell's Palsy?
Abdul Qader Ismail, BMSCH, Oluwasey Alake, MRCFCH, and Chetana Kallapps, MRCFCH

Conclusion – all children recovered, with or without steroids...further studies needed....

CAVE
Retrospective study – some treated, some not
No grading!
Newer large controlled trials in adults not even cited
Acute IX / X glossopharyngeal / vagus nerve palsy

- Exceptional!
- «Idiopathic-parainfectios» (post vaccination)
  (historic – diphtheria)
- Paresis of palate
  - if unilateral – may be asymptomatic
  - nasal voice (rhinolalia)
  - fluid regurgitation into nasal cavity
  - dysphagia
- Examination
  - failure to elevate palate
  - unilateral → deviation towards unaffected side

Acute hypoglossal nerve palsy

- Very rare!
- Problem – onset may not be realised
  (seen patient with XII nerve palsy as an «incidental» finding)

Reported
- Complication of bacterial meningitis
- Following dental treatment (controversial)

Hypoglossal Nerve Palsy
during Meningococcal Meningitis

Recovery, 5 months later
«Cranial polyneuropathy» - bilateral

- Very exceptional
- Consider
  - within GBS-spectrum

Additional references

- Cheng DR et al
  Recurrent 6th nerve palsy in a child following different vaccines
  BMC Infectious diseases 2012;12:105
- Jukes C
  Benign recurrent sixth nerve palsy in an infant
- Leiba H et al
  Prediction of the clinical outcome of cavernous sinus lesions in children
  Neuropediatrics 2013;44:191-98