Acute cranial nerve deficits

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Inclusion - Exclusion

• Pediatric focus
• Acute < 2-3 days
• Not considered – acute hearing loss
• «Acute» - sometimes a longstanding problem is only recently realized («pseudo-acute»)

Acute → «categories» ?

• Trauma
• Infection – inflammation - demyelination
• 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
- IIIrd nerve palsy, pupil mostly not spared (variable)
- Outcome – spontaneous recovery in a few weeks
- Treatment – wait and see (steroids?)
- Recurrences possible
- MRI – enhancement of thickened intracisternal portion of III nerve
- CSF (literature) normal

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**Problem with definitions...**

May not be recurrent, not painful

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1. Some data suggest that headache can develop up to 14 days prior to oculomotor paresis.
2. Treatment with corticosteroids is beneficial in some patients.

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**13.8 Tolosa-Hunt syndrome**

*Description*: Unilateral orbital or peribulbar pain associated with paresis of one or more of the IIId, IVth, and/or VIth cranial nerves caused by a granulomatous inflammation in the cavernous sinus, superior orbit, or orbit.

*Diagnostic criteria*: A. Unilateral orbital or peribulbar headache fulfilling criteria C.

B. Both of the following:

1. Granulomatous inflammation of the cavernous sinus, superior orbital fissure, or orbit, demonstrated by MRI or biopsy.
2. Presence of one or more of the following:
   - Paresis of one or more of the IIIrd, IVth, or VIth cranial nerves.
   - Evidence of cavernous sinus compression demonstrated by both of the following:
     1. Headache is (polarized to the granulomatous inflammation).
     2. Headache has preceded paresis of the IIIrd, IVth, or VIth cranial nerves.
     3. Symptomatic improvement of the pupil is occasionally observed.

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**Painful Ophthalmoplegia – Tolosa Hunt Syndrome - Cavernous sinus lesion**

- Not an aetiologial entity
- Initial cranial nerve dysfunction variable (isolated, combined) mostly III > VI...combinations
- MRI – high priority
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 trochlear nerve palsy
• Most common etiologies
  trauma
  congenital

Acute trigeminal nerve palsy
• «Never seen»
• (Herpes zoster ophthalmicus?)

  Chronic V nerve dysfunction (corneal lesion) in neurological syndromes
  • Congenital indifference to pain
  • Pontine Tegmental Cap Dysplasia
  • Gomez-Lopez-Hernandez Syndrome

Acute abducens nerve palsy

  Abducens nerve palsy

  Distinctive

  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

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**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»)

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**Acute facial nerve palsy**

- „peripheral“ paresis (lower motor neuron affected)
  - In praxis: 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

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**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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**Cerebellar** low grade glioma – obstructing fourth ventricle → hydrocephalus → increased ICP → papilledema

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A Germ Cell Tumor Masquerading as Bell Palsy

Yasemin Ozkale MD**, Ilman Erol MD**, Salam Yanco MD**

Pediatr Neurol 2013;85:109

MRI at presentation (age 2 months)

MRI 2 months later

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)

Facial nerve grading according to House, 1983, Laryngoscope

<table>
<thead>
<tr>
<th>Grade</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>I: Normal</td>
<td>Normal facial function in all cases</td>
</tr>
<tr>
<td>II: Mild dysfunction</td>
<td>Right weakness noticeable only on close inspection. At rest: normal symmetry and tone. Motion: some to normal movement of facial muscles, ability to close eye with minimal effort and slight asymmetry, no spontaneous contractions of facial muscles.</td>
</tr>
<tr>
<td>III: Moderate dysfunction</td>
<td>Obvious but non-disabling difference between two sides on functional impairment, noticeable but not severe continuity, contracture and facial asymmetry. At rest: normal symmetry and tone. Motion: slight to no movement of facial muscles, ability to close eye with minimal effort, obvious asymmetry, patients with obvious but not disabling weakness, contracture, or left facial asymmetry are Grade III regardless of the degree of weakness in the other face.</td>
</tr>
<tr>
<td>IV: Severe dysfunction</td>
<td>Obvious weakness and/or complete asymmetry. At rest: marked asymmetry and tone. Motion: no movement of facial muscles, loss of eye closure or oral movements, in patients with Bell's palsy and no other cause of facial weakness</td>
</tr>
<tr>
<td>V: Total paralysis</td>
<td>Loss of tone, asymmetry, no movements, no spasm, contracture, or facial weakness.</td>
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</tbody>
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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]
  [based on 2 large controlled studies]

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 (shown by MRI)
→ compression within bony structures

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)
- «oculoprotection» (eye drops, «artificial tears»)

Information to child/parents
Arrange short-term follow-up

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

Do Oral Steroids Aid Recovery in Children With Bell’s Palsy?

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 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

**Acute visual loss in children**

<table>
<thead>
<tr>
<th>Children</th>
<th>Adults</th>
</tr>
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<tbody>
<tr>
<td>• Optic neuritis</td>
<td>• Retinal (detachment, ischemia)</td>
</tr>
<tr>
<td>• Functional</td>
<td>• Optic neuropathies</td>
</tr>
<tr>
<td>• …</td>
<td>ischemic</td>
</tr>
<tr>
<td>Optic neuritis</td>
<td>inflammatory</td>
</tr>
<tr>
<td>- Papillitis</td>
<td>-</td>
</tr>
<tr>
<td>- Retrobulbar neuritis</td>
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**Visual loss - Examination**

- Visual acuity (both sides separately)
- Visual fields
- Pupillary reactions (? Relative Afferent Pupillary Defect)
- Ocular motility (limitation ? movements painful ?)
- Fundus
- Reduced contrast acuities – reduced color vision
Visual acuity testing – separate for each eye

If severe reduction of vision
→ Check finger counting

Swinging flash-light test

• Test for Relative Afferent Pupillary Defect (RAPD)
• Prerequisite: unilateral lesion, no efferent defect, no anisocoria
• Normal reaction: initial constriction of pupil
• RAPD: initial dilatation of pupil
• RAPD: typical for optic nerve (not retinal) disease

Optic neuritis Children versus Adults

• Often postinfectious
• Usually associated with disc swelling (papillitis)
• Commonly bilateral

Optic neuritis

• Retroorbital pain: may precede visual loss / 1-2 weeks duration
• (Headache)
• Blurring – fogging of vision
• Rapid - subacute significant visual loss (hours 7 – 10 days)
• Reduced contrast acuities (reduced color vision, dyschromatopsia)
• Variable visual field defects (central scotoma, peripheral..)
• Fundus: retrobulbar neuritis → normal
• papillitis → swollen (blurred) disc (mild peripapillary hemorrhages possible)
• RAPD (Relative Afferent Pupillary Defect)

CLINICAL DIAGNOSIS

Optic disc blurred - swollen («papilloedema») - simplified!

• Increased intracranial pressure („symptomatic - idiopathic“)
  Vision normal
• Inflammation («Papillitis»)
  Vision markedly reduced

Pseudopapilloedema
• Optic disc Drusen
  Vision normal
• Other variants
  Vision normal
• Varia (myelinated retinal fibres, hypermetropia..)

Optic disc blurred – normal visual acuity
MRI normal, CSF (opening pressure) normal
Optic neuritis

-- Postinfectious – days or weeks [viral, EBV, bacterial, toxoplasmosis...]
-- «idiopathic»

-- ON as first Acquired Demyelinating Syndrome (ADS)
  • Isolated ON
  • ON with anti-MOG antibodies
  • ON with AQP4-antibodies (NMO spectrum)
  • ON in ADEM (monophasic, recurrent)
  • ON → multiple sclerosis
  • ....

MOG antibodies reported in

• ADEM
• Multiphasic DEM (MDEM)
• ADEM followed by ON
• Isolated ON
• Recurrent ON
• NMO/D without AQP4-ab
• Transverse myelitis
• II MS

MOG antibodies (in children)

• MOG ab prevalent (> 1/3) in ADS
• MOG ab at presentation ~ 50% relapses
• MOG ab at onset → non-multiple sclerosis course
• [Adult ON, Optic Neuritis Treatment Trial (n=177) 1.7% +MOG]
• Disease phenotypes depend on age
  younger children – ADEM
  older children – ON, TM

Duignan et al. DMCN 2018 (n=237 ADS)
  • 64 % +MOG in ADEM (45/70)
  • 96 % +MOG in relapsing DEM
  • 41 % +MOG in ON (28/65)
  • 06 % +MOG in TN (3/50)

Clinical diagnosis of optic neuritis – and now?

Additional investigations to consider

• Neuroimaging
  MRI brain
  (MRI spine)
• CSF examination
• Antibodies anti-MOG
  Anti-Aquaporin-4 (AQP4) [NMO]

Treatment

• Steroids
  - iv
  - oral
• (referral to ophthalmologist)

Conclusion: bioequivalent doses of oral corticosteroids may be used as an alternative to IV corticosteroids

Effect of Treating Acute Optic Neuritis With Bioequivalent Oral vs Intravenous Corticosteroids
A Randomized Clinical Trial

Adults (> 18 years)  55 randomized patients
3 days treatment:
  • iv 1000 mg methylprednisolone
  • oral 1250 mg prednisone
  No taper

No significant group differences in recovery / outcome

Conclusion: bioequivalent doses of oral corticosteroids may be used as an alternative to iv corticosteroids

Imaging of Orbit – Optic nerve – of limited value (personal view)