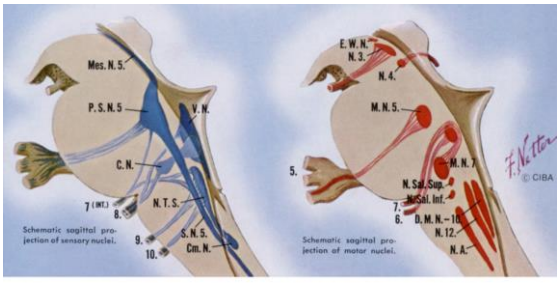
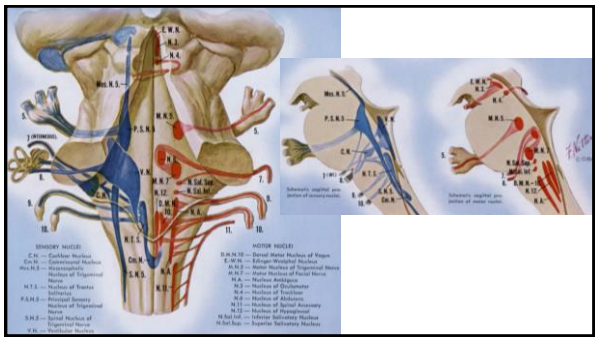
Acute cranial nerve deficits

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





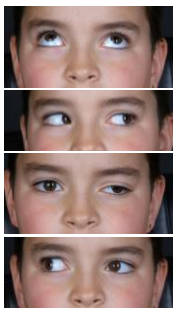
Acute → «categories» ?

- Trauma
- **Infection – inflammation**
- Haemorrhage

- Intracranial pressure
- Tumor
- Toxic

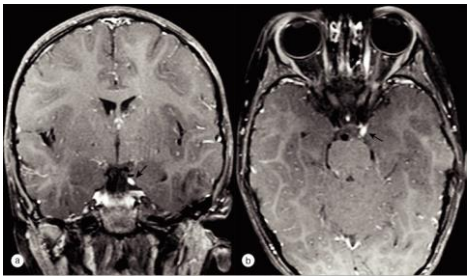
Acute oculomotor nerve palsy

Acute IIIrd nerve palsy

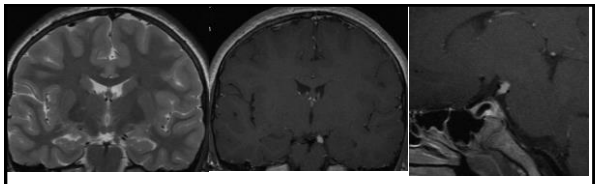


«Ophthalmoplegic migraine» (re-classified as cranial neuralgia)

- 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[20]:

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 acces 3rd ed – Cephalagia 2013;33(9)

ICHD-3 beta

Cephalalgia
International Headache Society
Cephalalgia
33(9) 629-808

DOI: 10.1177/0333102413485658
cep.sagepub.com
SAGE

Headache Classification Committee of the International Headache Society (IHS)

The International Classification of Headache Disorders, 3rd edition (beta version)

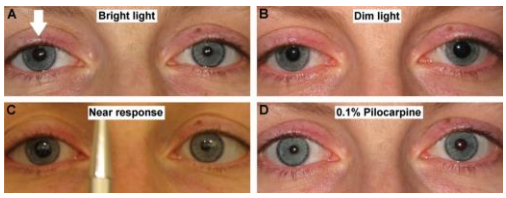
Acute (isolated) mydriasis

- 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


A
Bright light
B
Dim light
C
Near response
D
0.1% Pilocarpine

Acute abducens nerve palsy

Distinguish

Abducens nerve palsy — Abduction deficit

Non-localizing

Parainfectious, trauma, diabetes,
Increased intracranial pressure incl. Pseudotumor cerebri

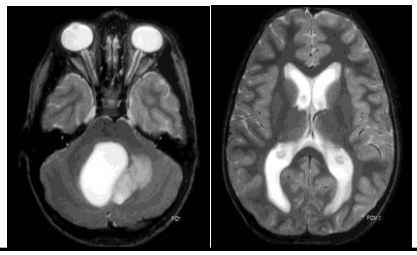
Localizing

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

Acute abducens palsy and *pontine glioma*

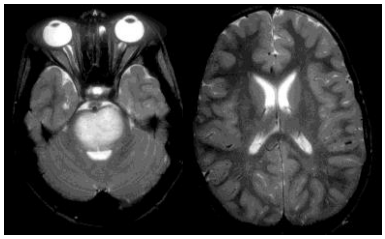
- Usually NOT isolated
  + other cranial nerve deficits and longtract signs

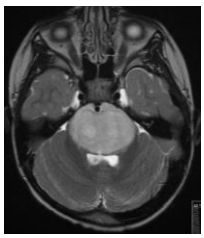
Cerebellar low grade glioma – obstructing fourth ventricle → hydrocephalus

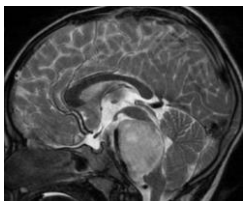


Intrinsic pontine glioma – fourth ventricle displaced, but patent

→ no hydrocephalus







Intrinsic pontine glioma

Acute benign abducens nerve palsy in infants

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

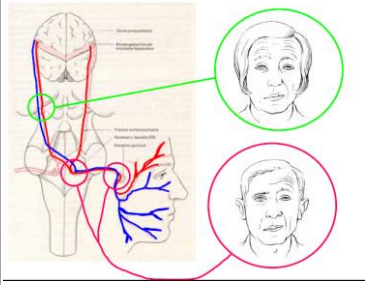
- Investigations – (a matter of «temperament»)

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

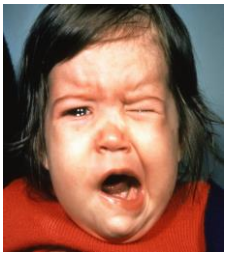
Red flags → further investiations
Paresis not isolated – subacute onset – bilateral
– central – Age < 2 years

Facial nerve paresis



Central
Forehead spared

Peripheral
All muscles affected



Right facial paresis

Hypoplasia M.depressor anguli oris right

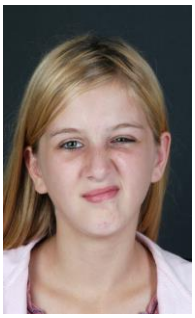
Acute peripheral facial palsy

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

Facial palsy
2nd «episode»

1st facial palsy 1 year ago



Melkersson-Rosenthal Syndrom

Cheilitis

Tongue changes not consistent
(Lingua plicata)

Clinical Picture NEJM June 29, 2013
Medical history – two episodes of peripheral nerve palsy

Persistent swollen lip: cheilitis granulomatosa

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 1 Facial nerve grading according to House [7]

Grade	Definition
I Normal	Normal facial function in all areas
II Mild dysfunction	Slight weakness noticeable only on close inspection. At rest: normal symmetry and tone. Motion: some to normal movement of forehead; ability to close eye with minimal effort and slight asymmetry. No synkinesis, contracture or hemifacial spasm.
III Moderate dysfunction	Obvious but not disfiguring difference between two sides; no functional impairment; noticeable but not severe synkinesis, contracture and/or hemifacial spasm. At rest: normal symmetry and tone. Motion: slight to no movement of forehead; ability to close eye with maximal effort and obvious asymmetry. Patients with obvious but not disfiguring synkinesis, contracture, and/or hemifacial spasm are Grade III regardless of the degree of motor activity.
IV Moderately severe dysfunction	Obvious weakness and/or disfiguring asymmetry. At rest: normal symmetry and tone. Motion: no movement of forehead; inability to close eye completely with maximal effort, asymmetrical movement of corners of mouth with maximal effort. Patients with synkinesis, mass action, and/or hemifacial spasm severe enough to interfere with function are Grade IV, regardless of degree of motor activity.
V Severe dysfunction	Only barely perceptible motion. At rest: possible asymmetry with droop of corner of mouth and decreased or absent nasolabial fold. Motion: no movement of forehead; incomplete closure of eye and only slight movement of lid with maximal effort; slight movement of corner of mouth. Synkinesis, contracture, and hemifacial spasm usually absent.
VI Total paralysis	Loss of tone; asymmetry; no motion; no synkinesis, contracture, or hemifacial spasm.

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

AAN guideline update 2012 [Neurology 2012;79:2209-13]
„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 Amercian Academy of Otolaryngology –Head and Neck Surgery Foundation

AAN recommendation ….**oral steroids should be offered**

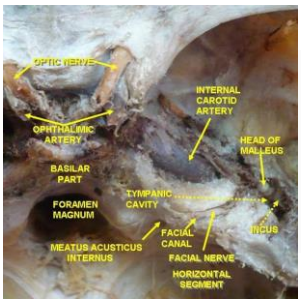
AAO-HNSF recommendation ….**clinicians should prescribe oral steroids**
within 72 hours of onset of symptoms

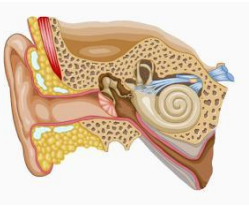
Rationale for steroid treatment

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





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»)

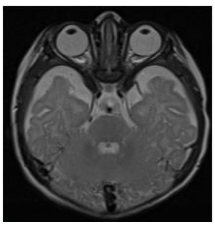
Information to child/parents

Arrange short-term follow-up

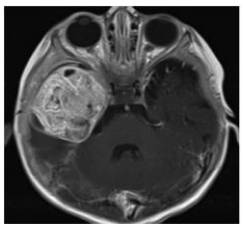
A Germ Cell Tumor Masquerading as Bell Palsy

Yasemin Özkale MD[a,*], Ilknur Erol MD[b], Nalan Yazıcı MD[c]

Pediatr Neurol 2013:49:509



MRI at presentation
(age 2 months)



MRI 2 months later

Original Article

Facial Nerve Paralysis in Children: Is It as Benign as Supposed?

Ariane Biebl MD*, Evelyn Lechner MD, Katarina Hroncek MD, Andrea Preisinger MD, Astrid Eisenkölbl MD, Klaus Schmitt MD, Dieter Furthner MD

Department of Pediatrics, Children's Hospital, Linz, Austria

Retrospective study, n=56

Pediatric Neurol 2014

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

J Child Neurol 2014;29:NP96
Authors from UK Birmingham

Abdul Qader Ismail, BMBCh[†], Oluwaseyi Alake, MRCPCH[†], and Chetana Kallappa, MRCPCH[†]

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 meningits
- Following dental treatment (controversial)

Hypoglossal Nerve Palsy during Meningococcal Meningitis

Recovery, 5 months later

IMAGE IN CLINICAL MEDICINE
Rockholt M, NEJM 2014;371

«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
  J Pediatr Ophthalmol & Strabismus 2014, e57
- Leiba H et al
  Prediction of the clinical outcome of cavernous sinus lesions in children
  Neuropediatrics 2013;44:191-98