OCULAR MOTILITY GRAND ROUNDS

Kelly A. Malloy, O.D., F.A.A.O.

ABSTRACT: Disturbances in ocular motility with resulting complaints of diplopia require prompt evaluation to correctly identify the disorder and determine etiology. This is a step-wise approach to ocular motility disorders, including CN III, IV and VI palsies, as well as brainstem motility disorders, focusing on identification and anatomic localization. A case-based approach demonstrates how an appropriate clinical evaluation and work-up is necessary to determine the correct etiology, and to ensure that the appropriate treatment is obtained.

"THE SIGNATURE OF CN III PARESIS"

- Hyper Deviation Which Increases In Upgaze And Reverses In Downgaze (Reversing Hyper Deviation), Exo Which Increases Across From The Vertically Limited Eye

DEATH FROM SUBARACHNOID HEMORRHAGE (SAH) (LOCKSLEY 1969)

- 20% Of Patients With SAH Die Within 48 Hours

ANSWER BY PUPIL

- INVOLVED=ANEURYSM - Posterior Communicating Artery (painful CNIII Palsy)
- SPARED=VASCULOPATHIC
- DOES NOT APPLY IF:
  -- Complicated CNIII, Incomplete CNIII, Relative Sparing, 20-50 Years Of Age

ANEURYSM Pathogenesis/Risk Factors

- Smoking/Drinking, Hypertension, Women >35 On Oral Contraceptives
- Family History, (polycystic kidney disease, aortic coarctation: NF-I)

ANEURYSM Treatment

- Medical-Vasospasm; Antifibrinolytics
- Surgical- Direct Clipping or Endovascular Occlusion-Coils

IS THIS REALLY A CN III PALSY? ANSWER BY PAIN

- ANEURYSM = 95%+
- DIABETES = 80%
- PITUITARY APOPLEXY, GIANT CELL, CAVERNOUS SINUS

"ANATOMICALLY GUIDED EXAM"

Fascicular (Midbrain)

- Contralateral hemiparesis (Weber’s), Contralateral hemiparesis / tremor of contralateral hand and foot (Benedickt’s), Contralateral ataxia (Claude’s)
CAVERNOUS SINUS
- III, IV, V1, V2, VI, Horner Syndrome, Proptosis (3mm.)

ABERRANT REGENERATION OF CN III
- Pseudo Graefe Sign, Eyelid Synkinesia, Light-Gaze Disassociation Pupils
  - COMMON CAUSES: Aneurysm, Tumor, Trauma
  - UNUSUAL: Inflammation, NEVER: Diabetes Mellitus

KIDS ARE DIFFERENT - ISOLATED CN III PALSY IN KIDS
- Congenital 44%, Trauma 16%, Inflammation 11%, Miscellaneous 11%
- Neoplasm 10%, Aneurysm 3%, Ischemia 3%

WHAT DO I DO FOR A PEDIATRIC CN III PARESIS?

CONGENITAL - MRI
ACQUIRED
- EXCLUDE TRAUMA OR MIGRAINE, CONSIDER LP (Lumbar Puncture) IF MRI IS NEGATIVE, CONSIDER MIGRAINE, IF MRI & LP ARE NEGATIVE > 10 years, ARTERIOGRAM

ISOLATED CN III PALSY IN ADULTS
- Undetermined 24%, Aneurysm 21%, Ischemia 18%
- Trauma 13%, Neoplasm 12%

CN III PALSY WHAT DO I DO? ADULTS
- 20-50 YEARS – CT, MRI, MRA, Arteriogram
- 50+ YEARS (pupil, palsy, pain) – Neuroimaging / Vasculopathic Evaluation

CN IV PALSY - ANSWER BY DIPLOPIA
- Vertical Diplopia, Worse at Near, Object Tilting, Relief With Head Position
- A Hypertropia That Increases Across From The Vertically Limited Eye And On Ipsilateral Head Tilt

MEASURING EXCYCLOTORSION
- Subjective, Maddox Rod, Bagolini Striated Lenses, Fundus, Plot Blindspot
OBJECTIVE vs. SUBJECTIVE EXCYCLOTORSION

- Objective = Subjective - Acute
- Objective > Subjective - Longstanding
- Objective Without Subjective - Cogenital

"CHECKLIST" EXAMINATION

what to look for: anatomic localization:

- CONTRA HORNER’S LOCUS CERULEUS (Copetto 1983)
- CONTRA INO MLF (Vannooteghem 1992)
- IPSI RAPD BRACHIUM SUP COLL (Elliot 1991)
- DMS ANT MEDULLARY VELUM
- BILATERAL CNIV ANT MEDULLARY VELUM
- TRUNCAL ATAXIA & IPSI DYSMETRIA SUP CEREBELLAR PEDUNCLE
- IPSI CN III, V1, VI, OSP CAVERNOUS SINUS

BILATERAL CNIV

- R HYPER> IN L GAZE & RHT, L HYPER> IN R GAZE & LHT
- V PATTERN ESO > 25 PD, EXCYCLOTORSION > 10 deg
- (LOOK FOR THE DORSAL MIDBRAIN SYNDROME)

ISOLATED CN IV IN KIDS (CAUSE)
- Trauma, Congenital

ISOLATED CN IV IN ADULTS
- 10% Aneurysm, 20% Ischemic, 30% Undetermined, 40% Trauma

TRAUMATIC CNIV PROGNOSIS (SYDNOR 1982)
- SPONTANEOUS RECOVERY:
  - UNILATERAL: 65%
  - BILATERAL: 25%

WHAT DO I DO? UNDER 20 YEARS
- History Of Trauma, Old Photos, Vertical Vergences, Dorsal Midbrain Syndrome?

WHAT DO I DO? 20-50 YEARS – Neuro-Imaging


CNIV: MANAGEMENT?
- Segmental Prisms & Patches X 9-12 Months
- Monitor For Secondary Contracture - Surgical Intervention
ABDUCTION DEFICIT/ CN VI PALSY

**CN VI PALSY KEY POINTS** - Measure At Infinity

- **Imposters – Abduction Deficits**
  - Graves Disease / Thyroid Orbitopathy, Myasthenia Gravis, Loss Of Fusional Reserves, Spasm Of The Near Reflex, Duane’s Retraction Syndrome

**“THE SIGNATURE OF CN VI PARESIS”**

- **At Distance:** Eso Which Increases In The Action Of The Palsied Eye

**CN VI PALSY - ANSWER BY MOTILITY**

- Duction > Version, “Glissades” (Slowed Saccades), Asymmetric OKN (Optokinetic Nystagmus), Negative Forced Duction

**ANATOMIC LOCALIZATION**

**FASICAL\N\N CN VI +**

- ContraLateral Hemiplegia = (Raymond’s)
- VII + ContraLateral Hemiplegia = (Millard-Gubler)
- Etiology: Infarction, Demyelination, Tumor

**SUBARACHNOID**

- Increased Intracranial Pressure (Papilledema), AICA Aneurysm, Subarachnoid Hemorrhage, Trauma, Meningitis, Clivus Tumor, Post Infectious, Neurosurgical

**PETROUS**

- Petrous Apex/ Mastoid Infection, Inferior Petrosal Sinus Infection, Petrous Bone Fracture, Trigeminal Schwannoma, Lumbar Puncture, Myelography, Spinal/ Epidural Anesthesia

**CAVERNOUS SINUS/ SOF**

- Aneurysm, Thrombosis, CCF (Carotid Cavernous Fistula), Dural AVM (Arterio-venous Malformation), Tumor, Tolossa-Hunt, Herpes Zoster

**WHAT DO I DO FOR ADULT CNVI PARESIS?**

- CBC (Complete Blood Count), BS (Blood Sugar) / HEMOGLOBIN A1c, LYME TITER, RPR/ FTA-ABS (tests for syphilis), ANA (Anti-Nuclear Antibody), ESR (Erythrocyte Sedimentation Rate), C-REACTIVE PROTEIN, PLATELETS, & HEMOGLOBIN (tests for Giant Cell Arteritis)

**WHAT DO I DO FOR A PEDIATRIC CNVI PARESIS?**

- Exclude Trauma, Lyme Titer, MRI With Gadolinium, Lumbar Puncture

**CHRONIC CNVI PALSY (= OR > 6 months) – Harbingers of Serious Intracranial Disease**
BRAINSTEM MOTILITY DISORDERS

DIFFERENTIAL DIAGNOSIS (vertical diplopia)
• CNIII NERVE PALSY
• CNIV NERVE PALSY
• MYASTHENIA GRAVIS
• GRAVES’ ORBITOPATHY
• SKEW DEVIATION

SKEW DEVIATION
• SUPERIOR MUSCLES INTORT & INFERIOR MUSCLES EXTORT
• RECTI = PRIMARY ELEVATORS
• OBLIQUES = PRIMARY CYCLOTORSIONALS
• SUPERIOR RECTUS & SUPERIOR OBLIQUE “DOUBLE CROSS”
• MED & LAT VN = YOKE MUSCLES, IR & SO
• SUPERIOR VN = YOKE MUSCLES, SR & IO

OCULAR SKEW TORSION SIGN
• SKEW DEVIATION
• OCULAR TORSION
• TILT OF SUBJECTIVE VISUAL VERTICAL
• + HEAD TILT

WHY THE HEAD TILT?*
• DIPLOPIA?, TORSION?, VESTIBULOCOLLIC REFLEX (VCR)?

SKEW DEVIATION LOCALIZATION  (Brandt & Dietrich)
• HYPERTROPIC, INTORTED EYE IS OPPOSITE TO THE LESION BELOW THE GRAVICEPTIVE PATHWAY CROSSING
• HYPERTROPIC, INTORTED EYE IS IPSILATERAL TO A LESION ABOVE THE GRAVICEPTIVE PATHWAY CROSSING TO INCajal
• DISCONJUGATE OCULAR TORSION IMPLIES A LOWER LEVEL LESION & CONJUGATE TORSION A HIGHER LEVEL LESION

DORSAL MIDBRAIN SYNDROME (SIGNS)
• TECTAL PUPILS
• UPGAZE PARESIS (DOWNGAZE PARESIS, OR BOTH)
• RETRACTION NYSTAGMUS
• EYELID RETRACTION

DORSAL MIDBRAIN SYNDROME  (lesser signs)
• CN IV Palsy
• Skew Deviation
• Pseudo CN IV Palsy
• Vergence Problems
• Square Wave Jerks
• Downbeat Nystagmus

Dorsal Midbrain Syndrome (symptoms)
• Diplopia
  —Skew, CN IV, convergence spasm, pseudo-abducens palsy
• Vergence Difficulties
• Headaches
• Transient Visual Obstructions

Dorsal Midbrain Syndrome (cause)
• Pineal Gland
• Midbrain
• Aqueduct
• Temporal Lobe
• Pinealoma
• Tumors, avm, III ventricle, infarct, MS, syphilis
• Stenosis
• Herniation

Internuclear Ophthalmoplegia (Smith & Cogan 1959)
Unilateral (INO)
• Vascular
• Older
• Males
• Apoplectic
• Skew (43%)
• Upright Gaze Nystagmus

Bilateral (Bino)
• Demyelinating
• Younger
• Males = Females
• Progressive
• Skew (13%)
• Upright Gaze Nystagmus

Internuclear Ophthalmoplegia (causes)
• HYDROCEPHALUS
• TUBERCULOSIS MENINGITIS
• PARANEOPLASTIC ENCEPHALOMYELITIS
• HIV-CMV ENCEPHALITIS
• HEAD TRAUMA
• DOXEPIN OVERDOSE
• SLE
• MIGRAINE
• SUPRATENTORIAL AVM
• INTRACRANIAL TUMOR

BRAINSTEM MOTILITY (SKEW / INO ) WORK-UP?
• NEURO-IMAGING
  – MRI with GAD
    • DEMYELINATION
    • INFARCT
    • MASS
  – CT SCAN
    • BLOOD
• LAB TESTING