Management of Childhood Nystagmus:
Knapp Lecturer Workshop

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Physiologic Nystagmus

- Optokinetic Nystagmus: rail road
- Vestibular nystagmus: Caloric /Positional
- End point nystagmus
- Voluntary nystagmus

Nystagmus is a rhythmic regular oscillation of the eyes. It may consist of alternating phases of a slow drift in one direction with a corrective quick "jerk" in the opposite direction, or of to and fro slow, sinusoidal, "pendular" oscillations.

Nystagmus in infancy

- Sensory Nystagmus
- Infantile Nystagmus syndrome: Manifest nystagmus
- Spasmus Nutans syndrome
- Fusional Maldevelopment Nystagmus syndrome: Manifest-Latent nystagmus
- Nystagmus blockage syndrome

Ocular Conditions with Nystagmus Associated with Poor vision

Leber congenital amaurosis
Rod monochromacy
Blue-cone monochromacy
Hereditary optic atrophy
Optic nerve hypoplasia
Ocular albinism
Congenital stationary night blindness
Congenital retinoschisis

Ocular Conditions with Nystagmus

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Ocular Conditions Associated with Nystagmus
Pendular Nystagmus

Conditions with Paradoxic Pupillary Reactions
- Congenital stationary night blindness
- Congenital achromatopsia
- Optic nerve hypoplasia
- Leber congenital amaurosis
- Best disease
- Albinism
- Retinitis pigmentosa

Infantile Nystagmus Syndrome, INS: Manifest nystagmus

Nystagmus examination
1. Anomalous head posture, AHP
2. Pendular /Jerk: R/L or up/down beating
3. Increasing in R/L gaze
4. Increasing on occlusion of R/L eye
5. Visual acuity: Binocular / Monocular
6. Associated strabismus
7. Associated ocular disease, albinism

ETDRS/ Snellen’s vision charting

Right eye
- 6/36
- 6/24
- 6/18

Left eye
- 6/24
- 6/36
- 6/60

Binocular vision
- 6/12
- 6/12
Measuring Anomalous head posture

Goniometer
or a protractor
and a scale

Face turns ARE Purposeful!

Examination of Nystagmus

Head posture
for distance
and near

Look for
associated
squint

Changing head posture:

Periodic alternating nystagmus

Four muscle supramaximal recession surgery

Documenting Nystagmus
ELECTRONYSTAGMOGRAPHY

Electronystagmography, ENG

Dextroversion
Primary position
Levoversion

AHP: Right sided face turn

Nystagmus types

• Jerk constant velocity slow phase: vestibular
• Pendular
• Jerk increasing velocity slow phase: Manifest/INS
• Jerk decreasing velocity sp: M-Latent/FMDNS

Manifest Latent Nystagmus (Fusional Maldevelopment)

Infantile Esotropia Nystagmus in Abduction, IENA, Ciancia

Preop IENA showing eso and Adduction preference and alternate face turn
Postop BE MR recession with posterior fixation
Spasmus nutans

High frequency (12-14Hz), asymmetric, dysconjugate, multiplanar-torsional, pendular
Exclude chiasmal glioma: do CT/MRI

Videonystagmography (VNG)

Left INO:
Right Abduction nystagmus  Left Underacting Adduction

Nonsurgical management

- Optimal prescription, use of tinted glasses
- Contact lenses
- Prisms: Base-out
- Auditory biofeedback
  (Sharma et al. J AAPOS, 2000)
- Acupuncture

Acupuncture in nystagmus
Needles used for acupuncture

Needle used in Index study

Acupuncture in Nystagmus

Acupuncture in nystagmus

Before needle 5mins 10mins

15mins 20mins After removal

MD thesis of Dr A Dhivya

Strategies for surgical management

- Shifting the eccentric null position: horizontal / vertical / oblique
- Artificial divergence: MR recession/PFixn
- Correcting associated strabismus
- Four horizontal muscle recession
- Hertle-Dell’Osso procedure

Nystagmus types

- Jerk constant velocity slow phase: vestibular
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Manifest Latent Nystagmus (Fusional Maldevelopment)
Acupuncture in nystagmus

Dhivya A, Sharma P, Dureja GP, Saxena R: unpublished data

Planning for Anderson-Kestenbaum

Face turn to left, Eyes in dextro version

RLR recession, LMR recession: Anderson procedure
RMR resection, LLR resection: Goto procedure

The surgery is usually planned at about 4.5 to 5 years age

This girl had wriggly eyes with best vision in left gaze. Next day after recessions of RMR and LLR the face turn reversed.

Planning for Kestenbaum

Face turn to left, Eyes in dextro version

RLR recession, LMR recession: Anderson procedure
RMR resection, LLR resection: Goto procedure

Nystagmus Surgery usually done at about 5 years age when the head posture has become stable

PAN: Four Horiz recti supramax recessions

Kestenbaum surgery

- Anderson: yoke muscle recession
- Goto: yoke muscle resection
- Kestenbaum four muscle recession- resection: all mm 5mm
- Parks: classic MR rec/res:5,6, LR rec/res:7,8
- Augmented Parks: 1:classic+40% for 30d,
  2: classic+60% for 45d
Augmented Anderson’s procedure

Recessions: MR 9mm, LR 12mm

**Table 1:**

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Pre-op</th>
<th>Post-op1</th>
<th>Post-op3 mo</th>
<th>p</th>
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</thead>
<tbody>
<tr>
<td>Face turn (degrees)</td>
<td>32.50</td>
<td>36.89</td>
<td>35.00</td>
<td>0.15</td>
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</table>

**Gupta, Sharma & Menon J AAPOS 2006**

**Figure:**

**Pre-op** vs **Post-op 3months**

**Graph:**

**Nystagmus Intensity**

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Pre-op</th>
<th>Post-op3 mths</th>
<th>p value</th>
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<tbody>
<tr>
<td>Nystagmus intensity</td>
<td>22.02</td>
<td>10.06</td>
<td>.006</td>
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</table>

**Gupta, Sharma & Menon J AAPOS 2006**
Eccentric null with strabismus

- If the eccentric null is in adduction
  - and there is an esodeviation
    - MR recession of the dominant eye (right eye)
  - and there is an exodeviation
    - MR recession of the dominant eye + R & R of the other eye.

Vertical AHP:
Chin up / eyes down

RIR recession
RSR resection
LIR recession
LSR resection

Vertical AHP:
Chin down
Eyes rolled up

- Surgery: B/L SR recess 8-9mm
  + IO recession and anteropositioning

Vertical AHP:
Chin up

- Parks: B/L IR recess 4mm for <25 d
  plus B/L SR resect 4mm for > 25 d
- Parks and Mitchell: B/L IR recess 8mm
  plus B/L SR resect 8mm for > 25 d
- Roberts et al: B/L IR recess 5mm
  plus SR resect 7mm
- Repka: B/L IR recess 6mm and 8mm

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Pre-op</th>
<th>Post-op 1wk</th>
<th>Post-op 3 mo</th>
<th>p value</th>
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<tbody>
<tr>
<td>Stereopsis arcsecs</td>
<td>110.00 ± 176.94</td>
<td>125.00 ± 179.92</td>
<td>75.00 ± 133.25</td>
<td>.205</td>
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Limitation of movements after Aug Andersons

<table>
<thead>
<tr>
<th></th>
<th>Nil/minimal (0-5 degrees)</th>
<th>Mild (6-10 degrees)</th>
<th>Moderate (11-20 degrees)</th>
<th>Severe (20-30 degrees)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Preop</td>
<td>12</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Postop 1wk</td>
<td>0</td>
<td>12</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Postop 3mths</td>
<td>4</td>
<td>8</td>
<td>0</td>
<td>0</td>
</tr>
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</table>

Gupta, Sharma & Menon J AAPOS 2006
Torticollis: head tilt
Oblique/Torsional Kestenbaum

Right head tilt: to do R sided extorsion
- Horiz: RLR up, RMR down, LMR up, LLR down (De Decker)
- Vert: RSR nasal, RIR temp
- LSR temp, LIR nasal (von Noorden)
- Obliques: operate on anterior fibres:
  - RSO & LI O recess, RIO & LSO adv + res

- Horizontal recti shifted vertically
  - De Decker
- Vertical recti shifted horizontally
  - von Noorden
- Oblique muscles: torsional anterior fibres: recess / advancement + resection

After RIO Ant half advancement 8mm & RSO ant half recession 8mm. Left eye had poor vision

Four muscle recessions
Supra-maximal recessions of four horizontal recti
Unpublished MD thesis of Jatin Ashar

Preoperative ENG

Postoperative ENG

Hertle-Dell’Osso procedure
Disinsertion and reinsertion of four horizontal recti
Unpublished MD thesis of Jatin Ashar

Preop ENG

Postop ENG
Artificial Divergence

- Spielmann
- Based on acceptance of BO prism
- BMR recess: 5-12 mm.
- Results: 26% of 75 cases reported success!
- 9/75 cases had unfusable exotropia

Medical treatment of Infantile Nystagmus

- Baclofen tried earlier in Acquired PAN, but not found effective in Cong PAN
- Gabapentin (GABA agonist/Glutamate antagonist thru NMDA) effective in Infantile nystagmus
  (Sherry et al British Journal of Ophthalmology 2006;90:839-843)

Conclusions

- Cases with definite eccentric null up to 30° respond very well to Aug Andersons
- For more than 40°, resections are added
- Cases with PAN: Bilateral Aug Anderson
- Cases with FMDN with Squint: Correct the squint
- IENA, Ciancia: MR rec with post fixation
- Cases with no definite null: Auditory biofeedback, Acupuncture, Four muscle supramax recession or Hertle- Dell’Osso procedure may be done.