Managing Nystagmus Without An Eccentric Null Position
2016 AAPOS Meeting Workshop: Management of Childhood Nystagmus

Richard W. Hertle, M.D.
The Children’s Vision Center, Akron Children’s Hospital Akron, OH
The Northeast Ohio Medical College, Rootstown, OH
I have personal financial relationships with:

- OXFORD UNIVERSITY PRESS
- Grant/Research Support – THE NATIONAL INSTITUTES OF HEALTH, THE NATIONAL EYE INSTITUTE, BETHESDA MD, USA
- Grant/Research Support - THE REBECCA S. CONSIDINE RESEARCH INSTITUTE, AKRON, OHIO, USA
- Shareholder and Patent Holder with RBG GROUP, LLC, USA
Acknowledgements

- Dongsheng Yang, PhD, Shanghai, China
- Louis F. Dell’Osso, PhD, Cleveland, OH
- Larry Abel, PhD, Melbourne, AU
- Jonathan Jacobs, PhD, Cleveland, OH
- Raymond Kraker, MPHS, Tampa, FLA
- Edmond F. Fitzgibbon, MD, Bethesda, MD
- Susan B. Mellow RN, MD, Bethesda, MD
- Mitra Maybodi, MD, Bethesda, MD
- Robert Williams, PhD, Memphis, TN
- David B. Granet, MD, La Jolla, CA
- Deanna Stevens, MD, Columbus, OH
- William Anninger, MD, Columbus, OH
- Vannessa M Hill, Columbus, OH
- Joel S. Schuman, MD, Pittsburgh, PA
- Hiroshi Ishikawa, PhD, Pittsburgh, PA
- Leah Reznick, MD, Pittsburgh, PA
- Mingshia Zhu, PhD, Pittsburgh, PA
- Kenneth Adams, DO, Albuquerque, NM
- Matthew Kaufman, MS, Pittsburgh, PA
- Eric Hald, Pittsburgh, PA
- Tara Cronin, MD, Pittsburgh, PA
- Ellen Mitchell, MD, Pittsburgh, PA
- Jai Jeng, Pittsburgh, PA

Ginger: Hudson, OH
Scout: Wibeaux, MT

- Kristen Carey, MD, Pittsburgh, PA
- Stephanie Knox, MD, Akron, OH
- Robert Burnstine, MD, Akron, OH
- Erin Benjamin, MD, Akron, OH
- Michael Jandt, Wibeaux, MT
- Jennifer Eaton, Akron, OH
- Steven Schmidt, PhD, Akron, OH
- Jeffery Dunmire, Akron, OH
Classification of Eye Movement Abnormalities and Strabismus - Nystagmus Types

1. Peripheral Vestibular Imbalance
   - Meniere, drug toxicity

2. Central Vestibular Imbalance
   - Downbeat, Upbeat, drug toxicity

3. Instability of Vestibular Mechanisms
   - PAN

4. Disorders of Visual Fixation
   - Vision Loss, SSN, drug toxicity

5. Disorders of Gaze Holding
   - GEN, ?APN, drug toxicity

6. Acquired Pendular Nystagmus
   - central myelin, oculopalatal, Whipple, drug toxicity

7. Saccadic Intrusions and Oscillations
   - SWJ, MSO, opsoclonus

8. Miscellaneous Eye Movements
   - SO Myokymia, OM neuromyotonia

9. Infantile Nystagmus Syndrome
   - “congenital,” “motor,” “sensory,” idiopathic, nystagmus blockage

10. Fusion Maldevelopment Nystagmus Syndrome
    - Latent, manifest latent, nystagmus blockage

11. Spasmus Nutans Syndrome
    - Without optic pathway glioma
    - With optic pathway glioma
| Disease Name | INFANTILE NYSTAGMUS SYNDROME (INS)  
[Old Congenital Nystagmus and “Motor and Sensory” Nystagmus] |
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Criteria</td>
<td>Infantile onset, ocular motor recordings show diagnostic (accelerating) slow phases</td>
</tr>
</tbody>
</table>
| Common Associated Findings | Conjugate, horizontal-torsional, increases with fixation attempt, progression from pendular to jerk, family history often positive, constant, conjugate, with or without associated sensory system deficits (e.g., albinism, achromatopsia), associated strabismus or refractive error, decreases with convergence, null and neutral zones present, associated head posture or head shaking, may exhibit a ”latent” component, “reversal” with OKN stimulus or (a)periodicity to the oscillation. Candidates on Chromosome X and 6  
May decrease with induced convergence, increased fusion, extraocular muscle surgery, contact lenses and sedation. |
| General Comments | Waveforms may change in early infancy, head posture usually evident by 4 years of age. Vision prognosis dependent on integrity of sensory system. |
Evaluation Techniques: Afferent System

- **Vision testing procedures**
  - Behavioral Vision Testing (acuity, color, stereo)
  - Visual Evoked Responses (flash, pattern, sweep)
  - Electoretinography (flash, pattern)
  - Contrast, Color and Visual Field Testing
  - SD-OCT
Eye Movement Recordings

- **Methods**
  - “Contact” electrooculography
  - Infrared reflectance
  - Remote Video
  - Scleral contact lens/magnetic search coils.
“It has been found that such operation not only may greatly lessen torticollis, but may also improve vision by lessening the nystagmus itself..”

4,355 patients with INS, 2006-2014

- Age 1.5 - 67 years (ave 14 years).
- 63% male.
- Follow up after surgery 9-23 mos (ave 11.1 mos).
- 68% of the patients had other eye disease.
- 61% had an associated systemic diagnosis.
- 33% oculocutaneous albinism.
- 62% anomalous head posture.
- 21% had a periodic or aperiodic component.
- 71% had strabismus.
- 71% had a significant refractive error.

<table>
<thead>
<tr>
<th>Operation Type</th>
<th>Description</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Operation 1</td>
<td>Horizontal Head Posture Alone</td>
<td>22</td>
</tr>
<tr>
<td>Operation 2</td>
<td>Chin Down Head Posture (+/- Strabismus)</td>
<td>16</td>
</tr>
<tr>
<td>Operation 3</td>
<td>Strabismus Alone</td>
<td>15</td>
</tr>
<tr>
<td>Operation 4</td>
<td>Horizontal Head Posture + Strabismus</td>
<td>10</td>
</tr>
<tr>
<td>Operation 5</td>
<td>Chin Up Head Posture (+/- Strabismus)</td>
<td>10</td>
</tr>
<tr>
<td>Operation 6</td>
<td>No Head Posture, Strabismus or Vergence Damping</td>
<td>9</td>
</tr>
<tr>
<td>Operation 7</td>
<td>Multiplanar Head Posture (+/- Strabismus)</td>
<td>7</td>
</tr>
<tr>
<td>Operation 8</td>
<td>Vergence Damping Alone (Artificial Divergence)</td>
<td>6</td>
</tr>
<tr>
<td>Operation 9</td>
<td>Torsional Head Posture Alone</td>
<td>5</td>
</tr>
</tbody>
</table>
OPERATION 3 – STRABISMUS ALONE

OPERATE ON ALL FOUR HORIZONTAL RECTI
SMALL DEVIATIONS BILATERAL RECESS + TENOTOMY
OR
LARGE DEVIATIONS BILATERAL RECESS + RESECT

15%
OPERATION 4 – HORIZONTAL HEAD POSTURE + STRABISMUS

I.E. - 30 ET + OD PREFERRED + FACE RIGHT

HEAD STRAIGHT WITH 50 BO OD
AND
EYES STRAIGHT WITH 20 BI OS

OD LATERAL RESECT + MEDIAL RECESS FOR 50 PRISM
AND
OS MEDIAL RESECT + LATERAL RECESS FOR 20 PRISM
OPERATION 6 – NO HEAD POSTURE-STRABISMUS-VERGENCE DAMPING

OPERATE ON ALL FOUR HORIZONTAL RECTI

TENOTOMY WITH REATTACHMENT

9%
OPERATION 8 – VERGENCE DAMPING (ARTIFICIAL DIVERGENCE)

OPERATE ON ALL FOUR HORIZONTAL RECTI

BILATERAL MEDIAL RECESS 3.0 mm
BILATERAL LATERAL RECTUS TENOTOMY AND REATTACH

6%
Conclusions – EOM Surgery and INS

• **Nine Operation Systematic Approach**

• **Beneficial Effects of Surgery on:**
  - Binocular Optotype Visual Acuity
  - Head Posture
  - Strabismus
  - Nystagmus (ANAF)

• **Independent of**
  - Age
  - Operation Subtype
  - Associated

• Ocular Diagnosis
• Systemic Diagnosis
Brain Response to Proprioceptive Disruption at Enthesis


Fackelmann K, et. Al., Histochemical characterisation of trigeminal neurons that innervate monkey extraocular muscles. Prog Brain Res. 2008;171:17-20