Skew Deviation & the OTR
Extra ‘steps’ and the old ‘new’ language of vertical and cyclovertical strabismus

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Objectives

1. Understand the current use of the terms “ocular tilt reaction” and “skew deviation”

2. Be familiar with the anatomic & physiologic substrate of ocular tilt & skew

3. Know how to distinguish between skew and isolated oblique palsies at the bedside, recognizing limitations in our understanding of cyclovertical ‘palsies’
Why Care About Skew?

- A 51-year-old woman complains of vertical diplopia for a day. Her exam reveals subtle vertical strabismus.
- A CT scan of the brain is normal.
- Is it just a 4\(^{th}\)? Can she go home?
- She then develops symptoms of nausea, hearing loss, ventilation problems (dyspnea) and somnolence.
MRI & Angio - Basilar Occlusion

Mennel, Clin Exp Ophth 2005
Overview

1. Definition & Brief History of “Skew”
2. Vertical & Torsional VOR
3. Normal & Pathologic OTR
4. Skew vs. Superior Oblique Oblique Palsy
5. Implications & Future Directions
Modern Definition of Skew & OTR
Modern Definition of Skew & OTR

1. Skew Deviation
   - An acquired vertical* strabismus resulting from imbalance in vestibular (balance system) inputs to the oculomotor system

2. Pathologic OTR – Ocular Tilt Reaction
   - The triad of skew deviation, binocular torsion (ocular counterroll), and head tilt
A Brief History of Skew & Cyclovertical Strabismus

A Tale of Two Disciplines
A Brief History of Skew

1. 1850+ – Latin terminology is purely descriptive (e.g. “strabiusmus sursoadductorius” to describe overaction in adduction); no implication of oblique dysfunction

2. 1871 – Nagel first to figure out cycloduction; forgotten

3. 1900 – Hoffman & Bielschowsky ‘discover’ and popularize head tilt to diagnose “cyclovertical palsy”

4. 1913 – ocular tilt discovered in animals; forgotten

5. 1926 – Brain describes head tilt & skew from ear disease (otitis interna)
A Brief History of Skew

1. **1958** – Parks labels & delineates elements of the “three-step test” for cyclovertical muscle ‘palsy’

2. **1960s** – ‘skew’ is vertical strabismus that ‘doesn’t fit’ three-step test, and implies central cause

3. **1975** – Keane publishes 100 cases, mostly posterior fossa disease, unclear localizing value; Westheimer & Blair (re)‘discover’ & characterize OTR in monkeys

4. **1977** – Rabinovitch et al. report 1st human OTR in MS

5. **1979** – Halmagyi et al. report 1st human OTR from peripheral vestibular lesion
A Brief History of Skew

1. 1980s – skew & OTR linked, but not linked to generic cyclovertical muscle ‘palsies’

2. 1991 – Brandt & Dieterich describe 3 ‘types’ of skew; later show localizing value with brainstem lesions

3. 1999 – Donahue et al. 5 cases of skew mimicking SO palsy, then 6 cases of skew mimicking IO palsy

4. 2001 – Demer points out tautologic thinking in strabismology of cyclovertical terminology

5. 2003 – Brodsky considers possible ‘horizontal skew’
Vertical & Torsional VOR
**Vertical & Torsional VOR**

**What is the V.O.R.?**

- The *vestibulo-ocular reflex*…
  1. Is the **circuit (reflex arc)** connecting balance organs and eye movement structures
  2. **Keeps our vision steady** when we move, by instantaneously moving our eyes in the direction opposite any head movement
Vestibulo-Ocular Reflex
Vertical & Torsional VOR

The Ear

- Outer Ear
- Middle Ear
- Inner Ear
- Balance Organ = labyrinth
Vertical & Torsional VOR

Inner Ear

Labyrinth

Semicircular Canals x 3
(A-VOR)

Otolith Organs
(L-VOR, OTR)
Vertical & Torsional VOR

Inner Ear to Brainstem

Labyrinth

Vestibular Nuclei

8th Nerve
Angular VOR Substrate

Vestibular nuclei (8th) are connected to the oculomotor nuclei

- **3rd & 4th** – to move the eyes vertically and torsionally
- **6th & 3rd** – to move the eyes horizontally
Vestibulocerebellum (the green colored stuff)

Vestibular Nuclei

Gaze-holding structures

8th Nerve
Vertical & Torsional VOR

How does the VOR work?

1. Towards the canal(s) is ‘ON’

2. Tonic firing at rest; rate changes with head motion; reciprocal innervation

3. Stimulates eyes to move in plane of canal, opposite head rotation
Normal & Pathologic

OTR
Ocular Tilt Response:
Normal Anatomy
Maintain a Stable Horizon

At least if you’re a horse!!!
Dynamic Roll VOR

Which muscles are these?

L-SR
L-SO

Where are these?

R-IR
R-IO

Left Tilt

3 - SR, IR, IO
4 - SO

Ant/Post SCC

MVN
Static Normal OTR

R-IR
R-IO
L-SR
L-SO

3 - SR, IR, IO
4 - SO

INC

Utricle
LVN

cerebellum partly suppresses these movements in normals
Ocular Tilt Response: Patho-anatomy
Pathologic OTR

- Dynamic roll-plane VOR (i.e. damage to semicircular canal afferents)
  - Torsional nystagmus with slow phase rolling towards the shoulder opposite the hypertropia (i.e. in the direction of the head tilt) plus skew

- Static roll-plane OTR (i.e. damage to utricular afferents or integrating centers)
  - Static ocular counterroll & skew
Pathologic Dynamic Roll VOR

The fast phase of nystagmus beats towards the hyper eye damage dynamic pathway from RIGHT ear in LEFT MLF.
Static Pathologic OTR

compensatory damage RIGHT ear pathway equivalent of LEFT head tilt

'wrong way' head tilt
Pathologic OTR
(Ocular Tilt Reaction)

- Triad
  - Head tilt
  - Vertical (skew) deviation
  - Ocular torsion

Note that eyes are opposite that expected for head tilt!!!

Modified from Rabinovitch, Arch Ophth 1977
Skew vs. SO Palsy
Ophthalmoscopy & Photography

normal fovea sits 2-7º below center of disc...

...so intorsion is easier to ‘see’ than extorsion
Pathologic OTR – Ocular Torsion

RIGHT head tilt, LEFT hyperdeviation, fundus torsion towards RIGHT shoulder

Mossman, Neurol 1997

EXcyclotorsion OD  INcyclotorsion OS
Pathologic OTR

- Profound, lasting head tilts/OTR
  - **Midbrain** – Interstitial nucleus of Cajal [INC] (contralesional head tilts)
  - **Lateral medulla** (ipsilesional head tilts)

- Transient head tilts/OTR
  - **Pons** – medial longitudinal fasciculus [MLF] (usually contralesional head tilts)
  - **Labyrinth/8th Nerve** – ipsilesional head tilts
Skew vs. SO Palsy
Skew Deviation vs. SO Palsy

- Parks 3-Step test with Bielschowsky Tilt Test
  1. Which eye is higher?
  2. Is it worst looking away from the side of the hyper?
  3. Is it worst with head tilted toward the hyper?
1. Which Eye Is Higher?

- Pathologic OTR with LEFT hyperdeviation
- LEFT 4\textsuperscript{th} Nerve Palsy
2. Worse Looking Right or Left?

- Pathologic OTR with LEFT hyperdeviation
- LEFT 4\textsuperscript{th} Nerve Palsy

Variable, Could be Right

Should be Right
Bielschowsky Head Tilt Test

3. Worse with Tilt Right or Left?

- Pathologic OTR with LEFT hyperdeviation
- LEFT 4th Nerve Palsy

Should be Left

Should be Left
Skew Deviation vs. SO Palsy

- Parks 3-Step test with Bielschowsky Tilt Test
  1. Which eye is higher?
  2. Is it worst looking away from the side of the hyper?
  3. Is it worst with head tilted toward the hyper?

- 4th Step
  4. Is it worst looking down?
4. Worse Looking Up or Down?

- Pathologic OTR with LEFT hyperdeviation
- LEFT 4th Nerve Palsy

Variable, Could be either

Down early, Up late
Skew Deviation vs. SO Palsy

- Parks 3-Step test with Bielschowsky Tilt Test
  1. Which eye is higher?
  2. Is it worst looking away from the side of the hyper?
  3. Is it worst with head tilted toward the hyper?
- 4th Step
  4. Is it worst looking down?
- 5th Step
  5. Is the fundus of the hyper eye excyclorotated?
5. Is Hyper Eye Excyclorotated?

- Pathologic OTR with LEFT hyperdeviation
- LEFT 4th Nerve Palsy

Should be Incyclo (?) Should be Excyclo
Skew Deviation vs. SO Palsy

- **Parks 3-Step test with Bielschowsky Tilt Test**
  1. Which eye is higher?
  2. Is it worst looking away from the side of the hyper?
  3. Is it worst with head tilted toward the hyper?

- **4th Step**
  4. Is it worst looking down?

- **5th Step**
  5. Is the fundus of the hyper eye excyclorotated?

- **More Steps? MRI ocular muscles & pulleys?**
Skew vs. SO Palsy

Other Features Suggesting Skew

- **Symptoms**
  - Dizziness or vertigo
  - Oscillopsia
  - Nausea/vomiting
  - Dysarthria/dysphagia
  - Clumsiness or poor coordination
  - Balance or gait problems (except due to loss of stereopsis)

- **Signs**
  - Impaired dynamic visual acuity
  - Head thrust sign
  - Breakdown of smooth pursuit tracking or VOR suppression
  - Saccadic dysmetria
  - Nystagmus
  - Limb or gait ataxia
Implication & Future Directions
Implications & Future Directions

1. Revisiting the ‘old’ language of cyclovertical strabismus (Demer, 2001)

2. ‘Secondary’ skews? Horizontal skews? (Brodsky, 2003); Pulley problems? (Demer)

3. Implications for strabismus surgery…
   1. Do the ‘opposite’ procedure? (Donahue)
   2. Will it work? (9/10 Siatkowski, beware cerebellar/ongoing)

4. Educational tools – NOVEL animations
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<thead>
<tr>
<th>Prevalent Conclusionary Term</th>
<th>Suggested Descriptive Term</th>
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<tbody>
<tr>
<td>Inferior oblique overaction</td>
<td>Over-elevation in adduction</td>
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Take Home Messages

1. “Skew” is more than strabismus that doesn’t fit SO palsy by 3-step test
2. Eye movement pattern in skew/OTR - towards ear is ‘on’; keeps horizon stable; ‘derivable’
3. Some skews look like oblique palsy except fundus torted the ‘wrong’ way
4. Ask about balance symptoms and look for signs in patients with vertical strabismus
5. Keep an open mind… this story is not over yet
ERRATUM 11/17/05

For those attending Moran Eye Institute Grand Rounds on 11/16/05, I answered a question after the lecture incorrectly. The correct answer is that direction of head tilt CANNOT distinguish between a superior oblique palsy and a pathologic OTR, since both will generally be ‘away’ from the hyperdeviated eye.