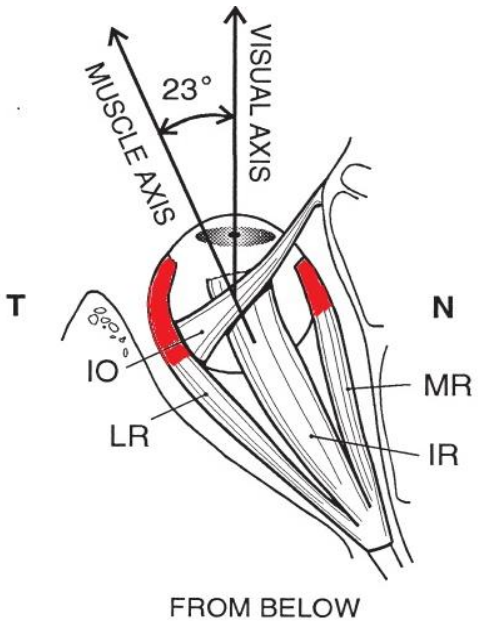
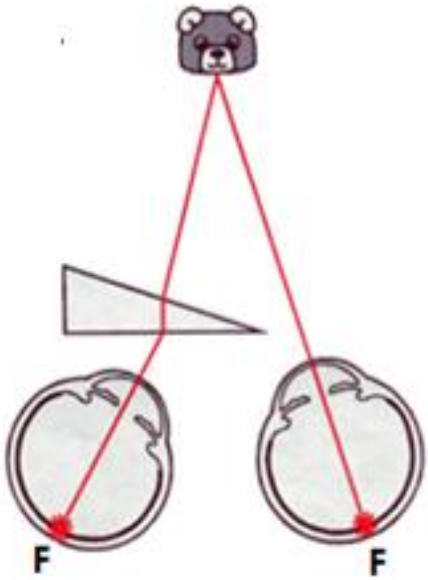
# Wright's Strabismus Review
# Crazy Easy

*Second Edition*


## By Kenneth W. Wright, M.D.



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Consultant and Editor


Wright Foundation

# Wright's Strabismus Review

## Strabismus Made Crazy Easy

*Second Edition*

### Preface First Edition

Strabismus is a topic often thought of as difficult. This book was written to simplify strabismus concepts and make them *Crazy Easy*. On one hand this is a board review book with questions interspersed throughout the text, yet it is also a complete and comprehensive strabismus text useful for the serious student of strabismus. I have included some corny jokes and weird comments in a weak attempt to make the lesson more fun. It is my sincere hope you find this text both informative and enjoyable.

Oh- Yes, that's me surfing in Panama after a terrific strabismus surgical mission.

Kenneth W. Wright, MD

### Preface Second Edition

I have been studying and working in the field of strabismus for over 30 years and continue to love my work. When your work is your hobby – you love your work. Strabismus affects our patients in many ways. They can lose vision from amblyopia, suffer from double vision, lose depth perception, and develop an inferiority complex because of the way people react to their appearance. Treating strabismus is very rewarding as in many patients it is life changing. The goal of this book is to present comprehensive review of strabismus in a clear and concise manner, emphasizing the understanding of fundamental principles. The book has questions interspersed throughout the text to help solidify principles and also to help you pass your boards. The most important goal of the book and the reason I wrote it is to help you help your patients with strabismus.

Respectfully,

Kenneth W. Wright, MD

# Wright's Strabismus Review

*Second Edition*

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# Wright's Strabismus Review
## Crazy Easy

*Second Edition*

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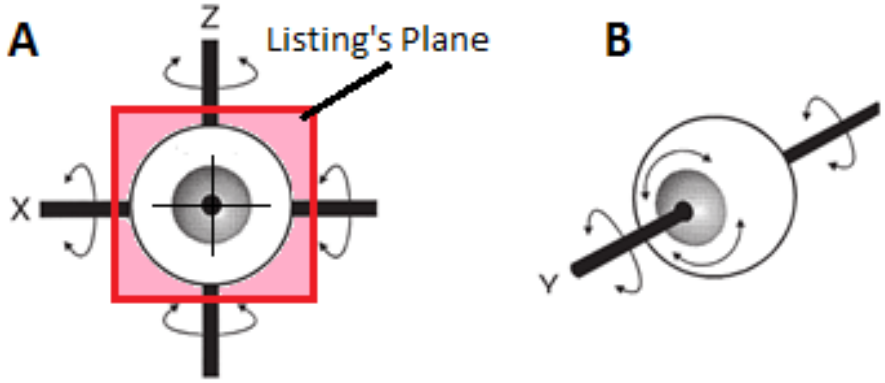
# Chapter 1
# Anatomy of Eye Movements

## Mechanics of Eye Movements

The eye is a sphere that rotates around three axes (see figure A and B):

A) Z axis for horizontal movements
B) X axis for vertical movements
C) Y axis for torsional movements

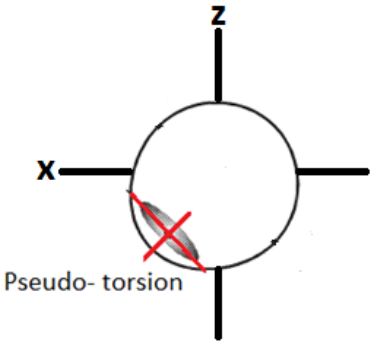
*Listing's plane* is a vertical plane that transects the X and Z axes and passes through the center of the eye (figure **A**).




True torsion occurs around the Y axis as shown in **B** above and is usually caused by oblique muscle overaction or under action.

***Listing's law***: All eye movements, (i.e., horizontal, vertical, and oblique) can be achieved by rotations around the X and Z axes. Oblique movements such as "up and out" or "down and in" combine horizontal (Z axes) and vertical (X axes) rotations on Listing's plane.

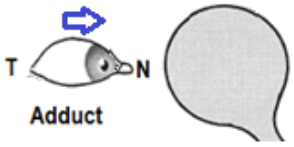
***Donder's law:*** Oblique rotations on Listing's plane which combine horizontal and vertical rotations will induce pseudo-torsion. Note the oblique rotation shown right on Listing's plane induces a twist in the red cross which is pseudo-torsion as it is not true rotation around the Y axis.



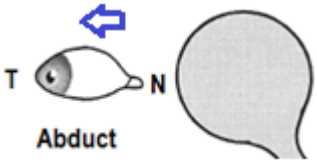
# Clinical Eye Movements

**Duction**: Ductions are monocular eye rotations and are evaluated by covering one eye and observing the range of motion of the fixing eye. Limited ductions indicate either restriction or muscle paresis or both. Versions are binocular eye movements that allows comparison of the movements of the two eyes (see P. 25).

## Horizontal Ductions

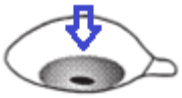



- Adduction – eye turns in to the nose (N) (to midline)

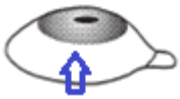



- Abduction – eye turns temporal (T) to the ear (away from midline)

## Vertical Ductions

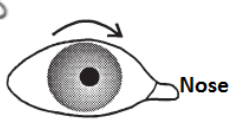



- Infraduction – eye moves down

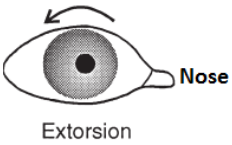



- Surpraduction – eye moves up

## Torsional Movements (a twisted movement)

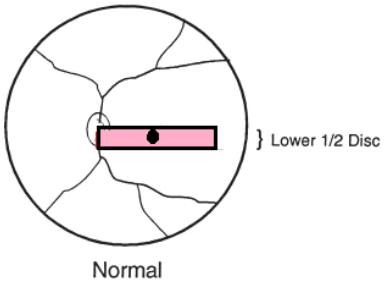



- Intorsion – 12:00 o'clock position rotates in to nose



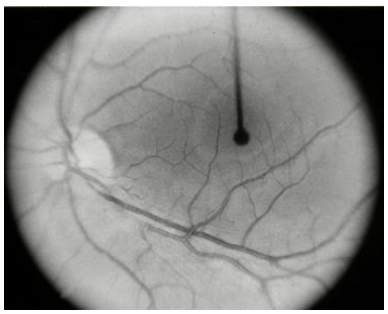

- Extorsion – 12:00 o'clock position rotates to ear

Ophthalmoscopy: The normal location of the fovea using direct ophthalmoscope is between the mid disc and inferior pole of the optic disc (see figure right) and indicates no significant torsion. If the fovea is higher than the center of the disc this is intorsion, and if the fovea is lower than the inferior pole of the optic disc this is extorsion.




**Question:** Fundus photo direct view right shows:

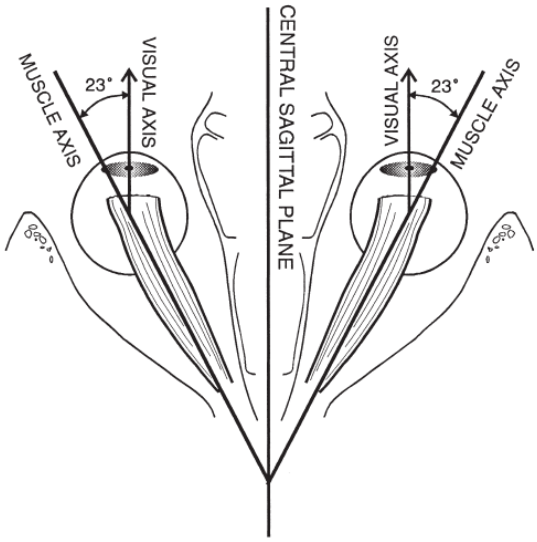
1. Intorsion
2. Extorsion
3. Normal – No torsion
4. Pseudo-torsion



**Answer:** This is a left eye and is intorted because the fovea is above the middle of the optic disc. The 12:00 o'clock position is rotated nasally so the answer is # 1.

## Anatomy of the Extraocular Muscles

The orbits and vertical rectus muscles are divergent being 23 degrees divergent from the central sagittal plane. The divergence of the orbits gives us a tendency for the eyes to drift out (exodeviation). When the extraocular muscles are relaxed such as during sleep, deep anesthesia, or when you are bored from reading this strabismus review the eyes will take a position of rest in the divergent position. Note that we keep our eyes on target by convergence that counters the natural tendency to diverge.




## Extraocular Muscle Fibers

Striated extraocular muscle fibers are made up of actin and myosin filaments which overlap to contract and shorten the muscle. Extraocular muscle fibers are more variable in size, are considerably smaller, and contract more than 10 times faster than other skeletal muscle. Extraocular muscle fibers are innervated at a high nerve fiber to muscle fiber ratio of almost 1:1, in contrast to other skeletal muscle that have up to 100 muscle fibers for every nerve fiber. This rich innervation, teamed with a fast muscle reaction time, contributes to the precision, accuracy, and control of eye movements. Another distinction of extraocular muscles is the presence of two muscle fiber types: fast muscle fibers and slow muscle fibers.

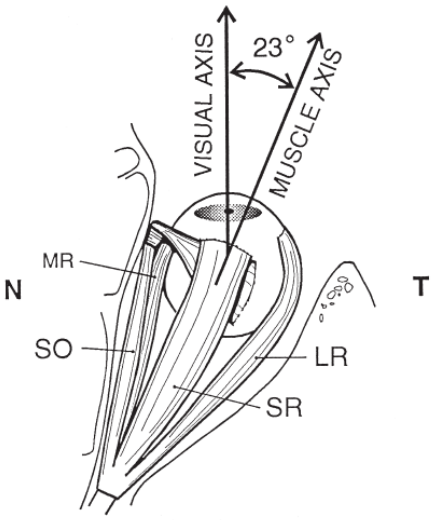
**Extraocular Muscle Fibers**

- **Fibrillenstruktur** - thin and fast
- **Felderstruktur** - slow or tonic

## Extraocular muscles

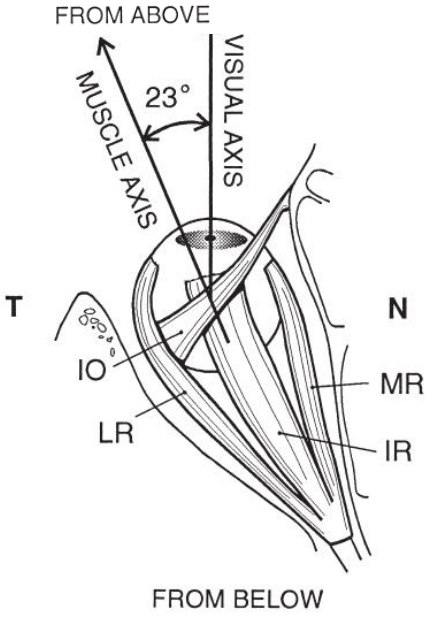
There are six extraocular muscles: four rectus muscles and two oblique muscles (see figures right view from above and below).

- **Four Rectus Muscles:**
  - Horizontal- medial, lateral
  - Vertical - superior, inferior
- **Two Oblique Muscles**:
  - Superior
  - inferior




***EOM Innervation:***

3rd Nerve -
Superior division – SR and Levator
Inferior Division – MR, IR, and IO
6th Nerve – LR
4th Nerve – SO

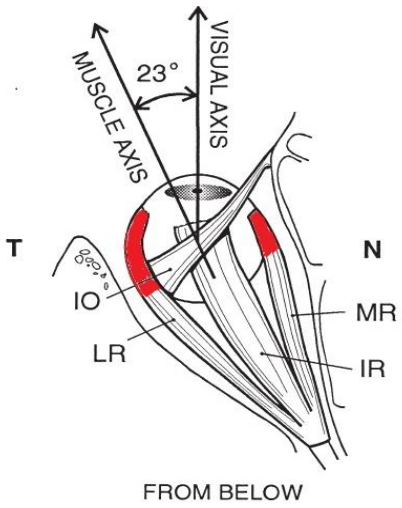



## Rectus Muscles:

The rectus muscles are 40 mm long. Rectus means straight and they course straight from the annulus of Zinn (apex of orbit) anteriorly to insert on sclera several millimeters from the limbus. The two vertical rectus muscles follow the orbit and approach the eye at a divergent angle of 23 degrees temporal to the visual axis (drawings right). Rectus muscles pull the eye posterior so a detached or lost rectus muscle will allow the eye to move forward and cause relative proptosis. A rectus muscle resection (muscle tightening procedure) can cause globe retraction producing relative enophthalmos. The nerves of the rectus muscles enter the muscle belly 2/3 posterior to the insertion or at approximately 15 mm from the annulus of Zinn.

## Arc of Contact

The arc of contact is the area of muscle that is in contact with the globe (see red in drawing right). Because of the divergence of the orbits, the lateral rectus has the longest arc of contact of the rectus muscles at 10 mm as it has to wrap temporally around the globe. The medial rectus has the shortest arc of contact at 6 mm. The oblique muscles have a longer arc of contact than the rectus muscles with the SO 12 mm and IO 15 mm.




## Question:

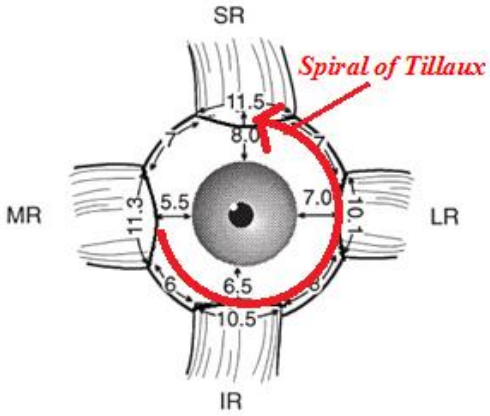
Which rectus muscle has the longest arc of contact to the globe?

1. Medial rectus
2. Lateral rectus
3. Inferior oblique
4. Superior oblique

**Answer:** The answer is # 2, lateral rectus muscle. Note the oblique muscles have a longer arc of contact but the question is which "rectus muscle" not which extraocular muscle - so be careful out there with board questions!

## *Insertion Notes:*

The medial rectus muscle insertion is closest (5.5 mm), and the superior rectus insertion is farthest (8.0 mm) from the limbus. This is the basis of the **spiral of Tillaux** which represents that the rectus muscle insertions are progressively farther from the limbus starting with the medial rectus. Note: the center of the insertion is closer to the limbus with the ends of the insertion (the poles) being more posterior thus giving a horseshoe shape insertion line.




- Remember the horseshoe insertions are galloping to the cornea.
- Note the muscle insertions are only 7 mm apart on average thus during strabismus surgery it is easy to hook the wrong muscle if not careful.
- Note the insertion width is about 10 mm.

**Question:**

When hooking the medial rectus muscle in surgery it is necessary to pass the hook posterior to the limbus by at least:

1. 3.5 mm
2. 4.5 mm
3. 5.5 mm
4. 7.0 mm

**Answer:** Because of the horseshoe configuration of the rectus muscle insertions, when hooking a rectus muscle one must pass the hook a few millimeters more posterior than the distance of the center of the insertion to the limbus (the standard numbers we memorize for the board exam). Since the limbal to center of insertion distance is 5.5 mm for the medial rectus to hook behind the insertion poles you have to pass the hook at least 7 mm so the best answer is # 4.

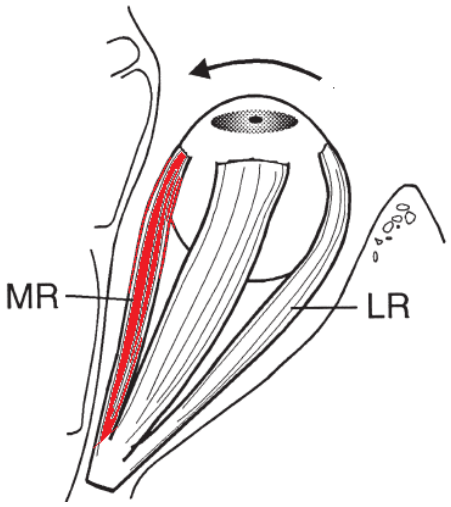
## Table of EOM Measurements

**TABLE 8-1. Extraocular Muscles.**

| *Muscle* | *Approximate muscle length (mm)* | *Origin* | *Anatomic insertion (mm)* | *Tendon length (mm)* | *Arc of contact (mm)* | *Action from primary position* |
|---|---|---|---|---|---|---|
| Medial rectus | 40 | Annulus of Zinn | 5.5 | 4 | 6 | Adduction |
| Lateral rectus | 40 | Annulus of Zinn | 7.0 | 8 | 10 | Abduction |
| Superior rectus | 40 | Annulus of Zinn | 8.0 | 6 | 6.5 | Elevation<br>Adduction<br>Intorsion |
| Inferior rectus | 40 | Annulus of Zinn | 6.5 | 7 | 7 | Depression<br>Adduction<br>Extorsion |
| Superior oblique | 32 | Orbit apex above annulus of Zinn | From temporal pole of superior rectus to within 6.5 mm of optic nerve | 26 | 12 | Intorsion<br>Depression<br>Abduction |
| Inferior oblique | 37 | Lacrimal fossa | Macular area | 1 | 15 | Extorsion<br>Elevation<br>Abduction |

## Medial Rectus Muscle (MR)

Action – Pure Adduction




The MR is the only extraocular muscle that has no attachment to other muscles. So the MR is most difficult to retrieve if inadvertently lost during surgery because it retracts posteriorly behind the globe.

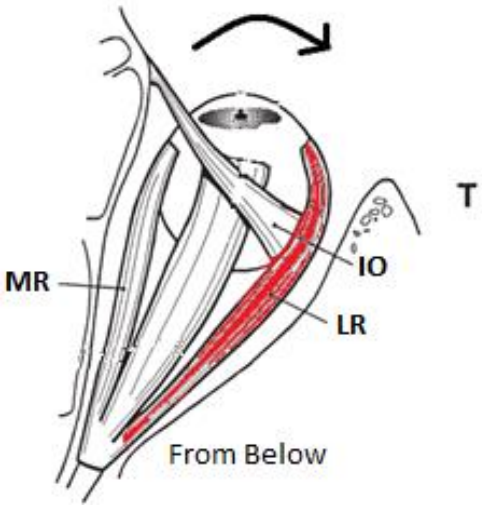
### *Medial Rectus - Three Shortest:*

- Shortest arc of contact (6 mm) of *rectus* muscles
- Shortest tendon (4 mm) of the rectus muscle *
- Shortest distance (5.5 mm) to limbus

*Note: The inferior oblique has the shortest tendon of all extraocular muscles at 1 mm, however, the MR has the shortest tendon of the rectus muscles. This distinction can be important for board questions.

## Lateral Rectus Muscle (LR)

Action – Pure abduction




- A ligament connects the LR muscle to inferior oblique muscle insertion. This is important as a lost LR can be found at the IO insertion.
- Longest arc of contact of the rectus muscles

## Question:

If lost during surgery which rectus muscle is most difficult to find?

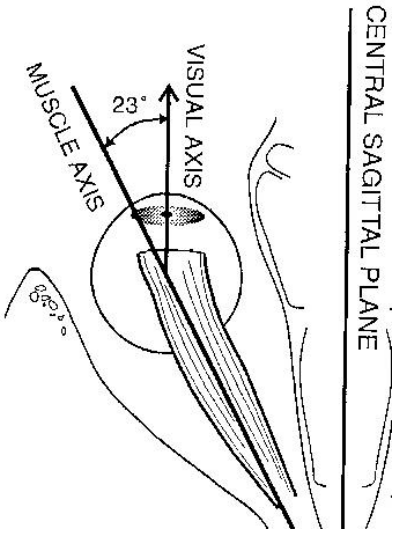
1. SR
2. IR
3. MR
4. LR

**Answer:** A lost MR muscle is most difficult to find so # 3 is correct. Since the MR has no attachment to other muscles, when lost it will retract posteriorly off the globe and end up in the orbital fat. The other rectus muscles have an attachment to an oblique muscle, so if detached they can be found attached to their oblique partner. When looking for a lost MR muscle look in the muscle sleeve in the orbital fat. Be careful not to follow the sclera posteriorly more than 10 mm from the MR insertion because the next structure you will find is the optic nerve.

## Vertical Rectus Muscles – Superior and Inferior

In contrast to the horizontal rectus muscles that have one action, the vertical recti have three actions:

- Primary action is vertical
  - SR - Elevator
  - IR - Depressor
  - Secondary action -
    - SR - Intorsion
    - IR - Extorsion
  - Tertiary action
    - SR and IR both - adduction



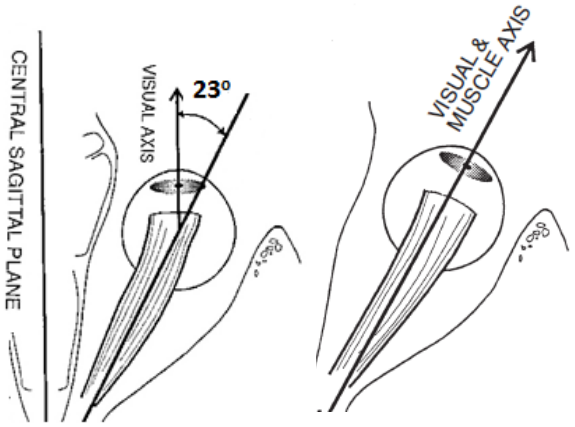

The discrepancy between the muscle axis and the visual angle is why vertical rectus muscles have more than one function. The vertical rectus muscle axis is 23 degrees temporal to the visual axis (see drawing right). Note: when the eye is abducted 23 degrees the muscle axis and visual axis are aligned and the vertical recti have one function: inferior rectus – depression and superior rectus – elevation.

### Question:

When do the vertical rectus muscles have a single vertical function?

1. The eye adducts 23 degrees
2. The eye abducts 23 degrees
3. In downgaze
4. The vertical rectus muscles always have secondary and tertiary functions.

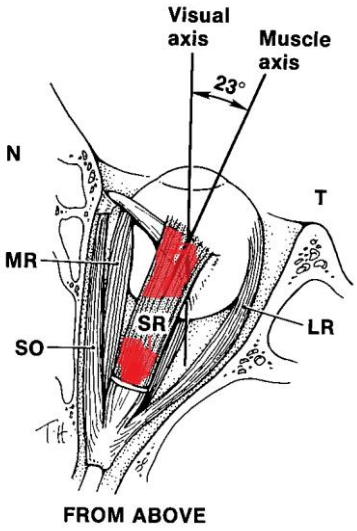
**Answer:** When the eye abducts 23 degrees the vertical muscle axis and visual axis will be aligned and the vertical rectus muscles then have one vertical function (diagrams right). The answer is # 2.




## Superior Rectus Muscle (SR)

Actions: Primary - elevation, Secondary - intorsion , Tertiary - adduction

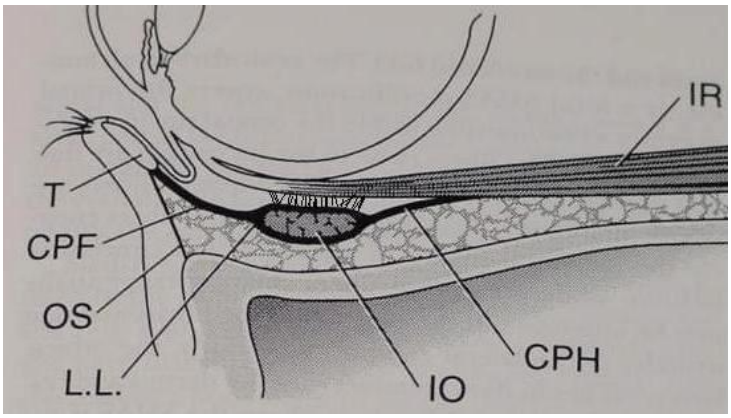
The superior rectus muscle is connected to superior oblique muscle below and levator muscle above (drawing right). Recession of SR can cause upper lid retraction as the upper lid moves posteriorly with the superior rectus muscle.



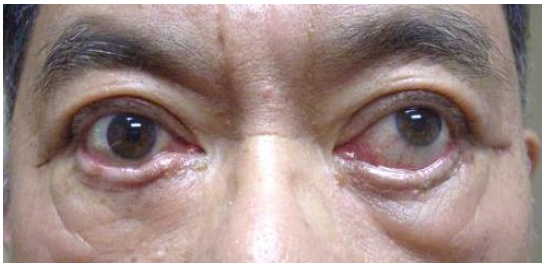

## Inferior Rectus Muscle (IR)

Actions: Primary - depression, Secondary – extorsion Tertiary - adduction

The IR muscle is connected to inferior oblique muscle below by Lockwood's ligament (drawing right). Recession or slippage of IR results in lower lid retraction. Find a lost IR muscle by finding the IO muscle as the IR will be attached to the IO muscle by Lockwood's ligament.




T= tarsus, CPF= Capsulopalpebral fascia, OS= orbital septum, LL= Lockwood's ligament. IO= inferior oblique, CPH= capsule-palpebral head, IR= inferior rectus



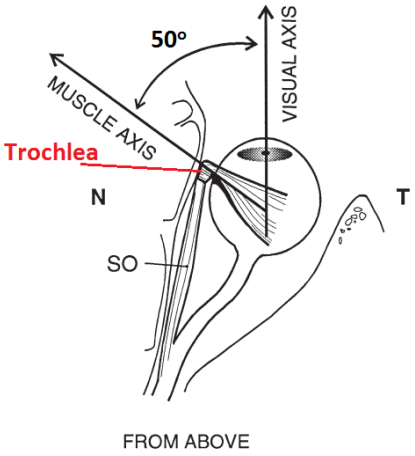
**Question:** Patient in photo right had bilateral lower lid blepharoplasty with a complication left eye. Which best describes why the left eye is up and out, and the lower lid is retracted?

1. Scarring of inferior orbital fat
2. Scarring for the lower lid skin incision
3. Traumatic disinsertion of IR muscle – lost muscle
4. Transection of SR muscle

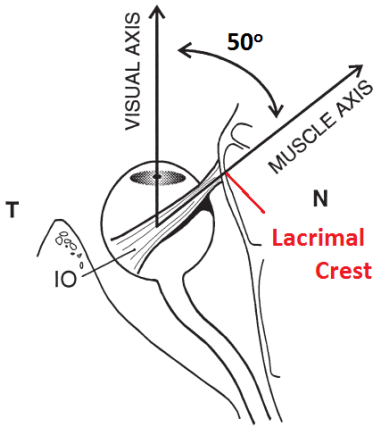
**Answer:** This patient had the surgical complication of disinsertion of the left IR muscle with posterior retraction of the IR muscle. The answer is # 3. The IR muscle is a depressor and adductor, so if the IR is disinserted the eye goes up and out. Because the IR is connected to the lower lid by Lockwood's ligament the lower lid retracts as the disinserted IR muscle retracts posteriorly. Photo right shows patient after your author retrieved and advanced the lost IR muscle. Note improvement of eye and lid position.

## Oblique Muscles: (Inferior and Superior Oblique)

- Primary action is torsion – SO intorsion, IO extorsion
- Secondary action is vertical
  - SO - depression
  - IO - elevation
- Tertiary action – Both abduction



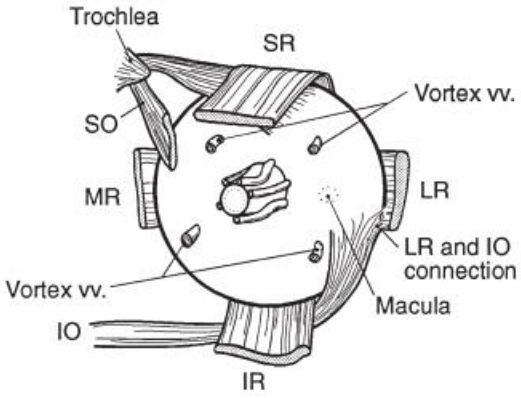

FROM ABOVE




FROM BELOW

Oblique muscles have an axis that is 50 degrees nasal to the visual axis which gives the obliques torsion as their primary action. The SO intorts as the top of the eye is pulled nasal, while the IO extorts as the bottom of the eye is pulled nasal (see drawings right). The posterior temporal insertion of the oblique muscles is why they have seemingly paradoxical vertical functions with the SO being a depressor and IO an elevator. The SO pulls the back of the eye in and up to the trochlea so the front of the eye goes down and out. The IO pulls the back of the eye down and in towards the lacrimal crest, thus rotating the front of the eye up and out.

## Oblique Notes: (see figure right)




- Oblique muscles underlie their vertical rectus partners.
- SO tendon posterior fibers insert close to the optic nerve (6 mm).
- IO muscle inserts close to the inferior temporal vortex vein and macula.
- The SO has the longest tendon (26 mm) and the IO the shortest tendon (1 mm) and that is generous!

### Question:

Which extraocular muscle group has the longest and shortest tendons?

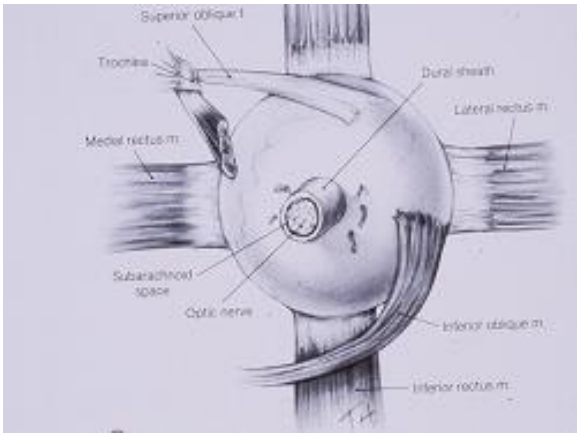
1. Horizontal rectus muscles
2. Vertical rectus muscles
3. Oblique muscles
4. None of the above

**Answer**: Longest Tendon = Superior Oblique – 26 mm, and the shortest tendon is the inferior oblique – 1 mm, so the answer is # 3, the oblique muscles. Note medial rectus has the shortest tendon of the rectus muscles.

### Question:

What is wrong with the drawing right from a popular anatomy textbook? (BTW - not one of my books)?

1. Inferior oblique muscle should be under the inferior rectus muscle.
2. SO insertion should extend posteriorly close to the optic nerve.
3. Both 1 and 2
4. The drawing is fine – It was published in a textbook so it has to be correct!




*Permission by Timothy Hengst, MA*

**Answer:** The correct answer is # 3. The SO tendon insertion is drawn too small. It actually has a broad insertion that extends from the superior rectus insertion to 6 mm from the optic nerve. Also the drawing shows the IO muscle between the sclera and the IR muscle. The IO muscle should be under the IR muscle like the drawing at the top of the page.

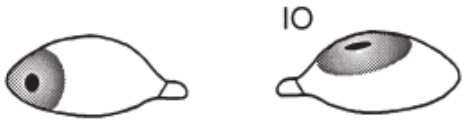
***Remember*:** The oblique muscles are a little odd – the superior is a depressor, and the inferior is an elevator, and they underlie their rectus partners.

## Inferior Oblique Muscle (IO)

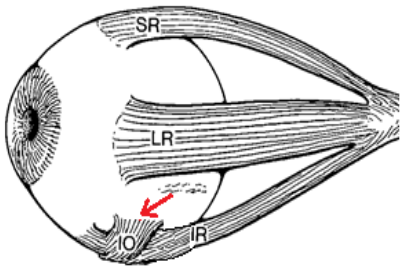
Actions: Primary - extorsion, Secondary - elevation, Tertiary - abduction

The origin of the inferior oblique muscle is the lacrimal crest located at the inferior anterior nasal aspect of the orbit. The IO muscle courses posterior temporally to insert behind the eye near the macula. When operating on the IO muscle watch out for the inferior temporal vortex vein as it is close to the IO insertion.

Inferior oblique muscle overaction is a common type of strabismus. It causes extorsion, up-shoot (shown in figure right), and abduction.

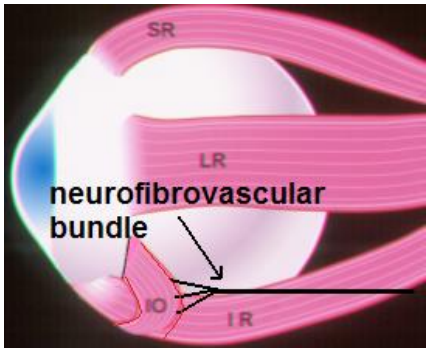



**Inferior oblique surgery:** Inferior oblique overaction can be reduced by surgically moving the IO insertion nasally towards it's origin thus slackening the muscle called an ***IO recession***. Also moving the IO insertion anterior towards the equator changes the IO from an elevator to more of a depressor and this surgery is called ***IO anterior transposition***.




**Note:** The ciliary nerve courses with the IO nerve. A deep posterior hooking of the IO muscle during surgery can traumatize the ciliary nerve thus causing the complication of pupil dilatation and reduced accommodation. Avoid this by using direct visualization of the posterior border of the IO muscle and hook right at the posterior border of the IO muscle. Avoid "deep blind" posterior passing of the hook which can traumatize the vortex vein and ciliary nerve.

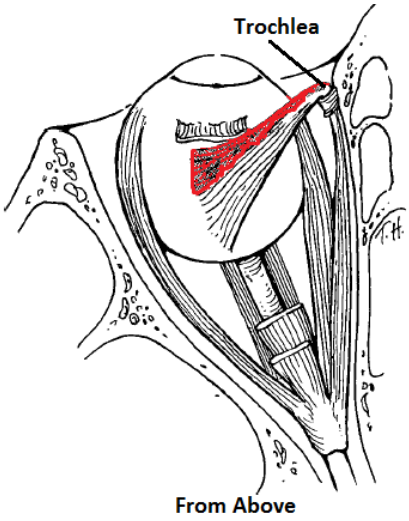
**Neurovascular Bundle:** A neurovascular bundle attaches to the posterior aspect of the IO muscle. If the posterior fibers of the IO muscle are anteriorized to the level of the inferior rectus insertion this will stretch the neurovascular bundle and cause a "J" deformity of the IO muscle (see drawing right). The tight neurovascular bundle attached to the IO muscle will pull the eye down and limit elevation (anti-elevation). A better surgical technique is to keep the posterior muscle fibers posterior to avoid anti-elevation, unless some limitation of elevation is desired such as the case of treating dissociated vertical deviation (see chapter 7 p 88).




## Superior Oblique Muscle (SO)

Actions: Primary - intorsion, Secondary - depression, Tertiary - abduction

The SO tendon is the longest (26 mm) of the extraocular muscles and it courses from its **anatomical origin** above the annulus of Zinn through the trochlea to insert broadly on sclera along the temporal border of the superior rectus muscle. The **functional origin** of the SO muscle is the trochlea, not the annulus of Zinn. Anterior tendon fibers (in red) are responsible for intorsion, while the posterior tendon (white and black) produces depression and abduction (see diagram right).




The SO muscle and its nerve (fourth nerve) courses outside the rectus muscle cone. This is why a retrobulbar injection of anesthetic will typically not anesthetize the SO muscle. Often the only movement after retrobulbar anesthesia is slight depression, slight abduction and significant intorsion as the SO is the only extraocular muscle still functioning.

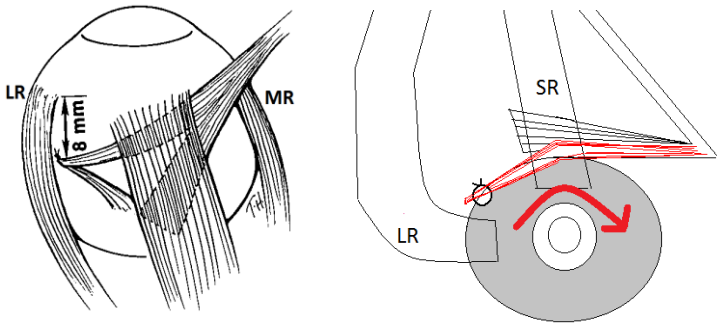
The superior oblique nerves exit the midbrain posteriorly close together thus closed head trauma that causes a shift in the brain will often result in a bilateral superior oblique palsy.

## Trochlea

The trochlea is a "pulley" composed of cartilage that redirects the SO tendon posteriorly. It is the functional origin of the SO muscle as the vector of SO muscle force pulls towards the trochlea. The superior oblique tendon is connected to surrounding cartilage by connective tissue. The muscle to tendon transition occurs in the area of the trochlea. The photo right shows a cross section through the trochlea and shows the cartilage that surrounds the muscle-tendon (T=tendon).


## *Harada-Ito Surgery*

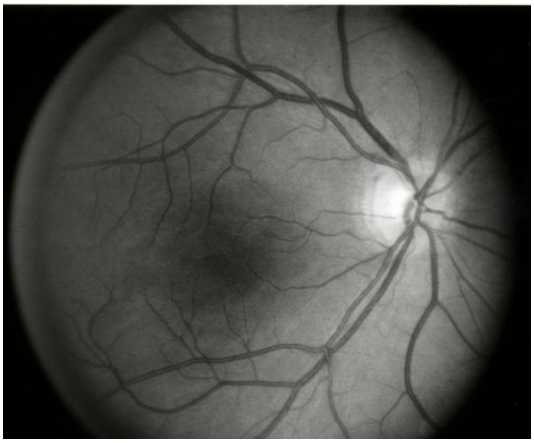
- Harada-Ito procedure tightens the anterior tendon fibers thus intorting the eye. Note the diagrams right show the anterior SO tendon fibers are pulled temporally towards the lateral rectus muscle thus producing intorsion.
- Harada-Ito is used to correct extorsion such as seen with a superior oblique palsy.




## Question:

Fundus photograph direct view of right eye in a patient with strabismus.

A. Which muscle could be weak?
   1. SO
   2. IO
   3. IR
   4. None of the above



B. Which surgical procedure would help correct this strabismus?
   1. SR recession OD
   2. IO recession OD
   3. SO Harada Ito OD
   4. # 2 and # 3

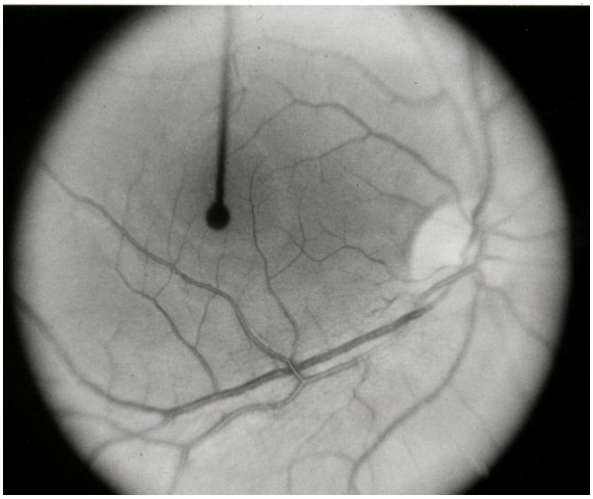
**Answer:**

A) This is a right eye. The fovea is below the inferior pole of the optic disc indicating significant extorsion. Since the SO muscle causes intorsion, a weak SO muscle would cause extorsion. The answer is # 1 SO. Note the IO and IR muscles are extorters so if they are weak intorsion would occur.

B) Extorsion can be corrected by strengthening the anterior fibers of the SO tendon i.e. the Harada-Ito procedure or by weakening the IO with an IO recession so the answer is # 4.

**Question:** Fundus photo direct view on the right suggests which muscle or muscles are overacting? (Could be more than one answer)

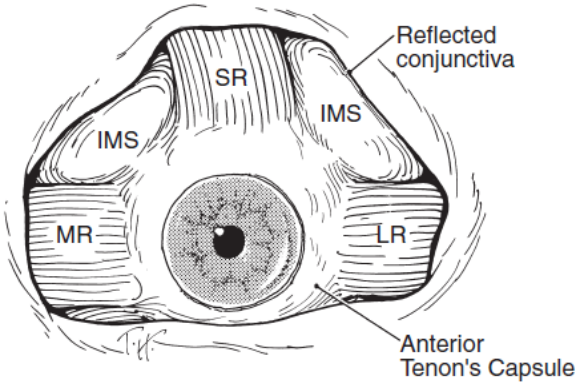
1. I.O.
2. S.O.
3. I.R.
4. S.R.



**Answer:** This is a right eye. The fovea is above the middle of the optic disc which indicates significant intorsion. Both the SO and SR muscles are intorters, so if they overact this would induce intorsion. The answer is # 2 and # 4. Note the IO and IR are extorter muscles.

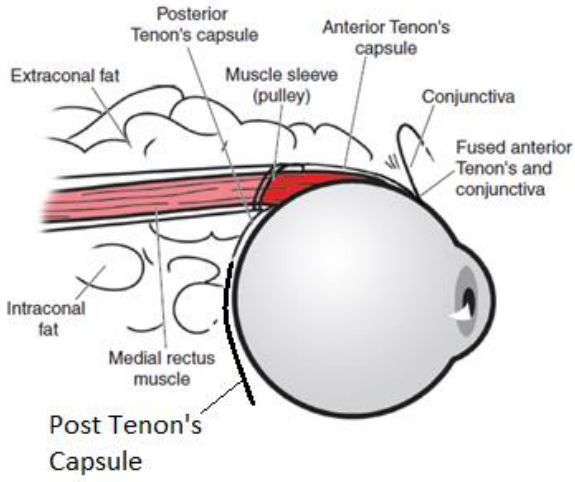
## Periocular Fascia and Tenon's Capsule

Tenon's capsule underlies the conjunctiva. It is a connective tissue that covers the sclera, muscles, and separates the eye from orbital fat. The elastic nature of Tenon's capsule allows the eye to rotate freely. Tenon's capsule is classified by its location. Intermuscular septum (IMS) is the tissue between the anterior aspect of the rectus muscles (see drawing right). Anterior Tenon's capsule is between the limbus and rectus insertions. At the limbus, Tenon's fuses with conjunctiva for 3 mm. Because the conjunctiva and anterior Tenon's capsule are fused at the limbus, a limbal incision will cut through both tissues and gain access to sclera with a single cut. Fornix incision requires two cuts to gain access to sclera, one through conjunctiva and a second through intermuscular septum. Anterior Tenon's capsule and intermuscular septum can be safely cut during strabismus surgery to gain access to the sclera and EOMs without complication.



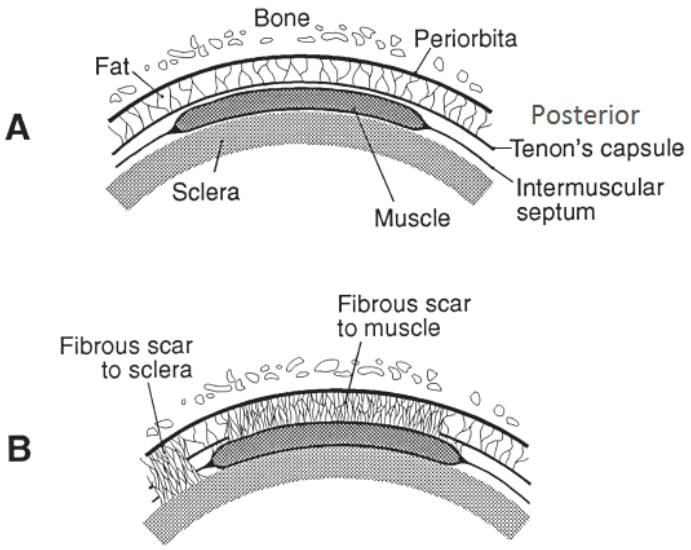

**Posterior Tenon's capsule** extends posterior to the rectus muscle insertions and separates orbital fat from the sclera thus allowing the eye to rotate freely (see drawing below). In contrast to anterior Tenon's capsule, posterior Tenon's should not be cut or violated during surgery as this can cause orbital fat to scar to the sclera (fat adherence ) causing restriction of eye movements (see below).

**Muscle sleeve** (AKA muscle pulley) is contiguous with posterior Tenon's capsule and is a hammock around the muscle that supports and controls the muscle trajectory to the eye. It is also a fascial barrier that keeps orbital fat posterior to the muscle insertion. Violation of the muscle sleeve can result in exposure of orbital fat and scarring of fat to the muscle or sclera (fat adherence) causing restriction of eye movement.



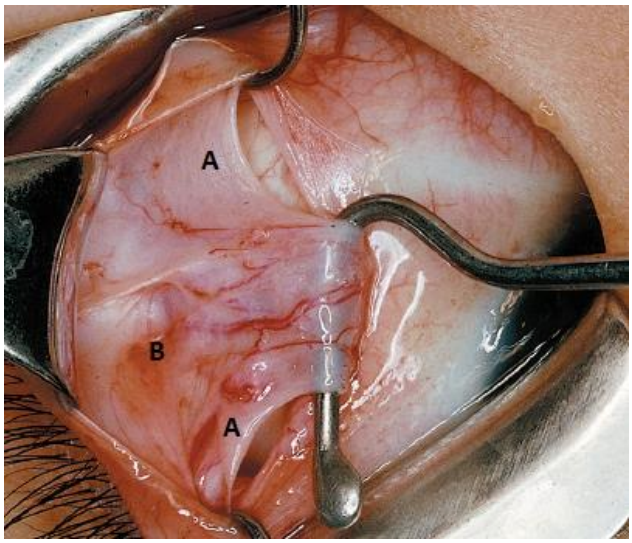
## Fat Adherence Syndrome

Crossectional drawing "A" shows normal relationship between sclera, muscle, and fat. Note that posterior Tenon's capsule separates fat from the muscle and sclera. Rupture of posterior Tenon's capsule will expose fat and casue fat adherence to muscle or sclera. Penetration of muscle sleeve AKA pulley, can also expose and traumatize fat and cause fat adherence to the muscle. Drawing "B" shows scarring and fat fibrosis to sclera and muscle. These adhesions will restrict ocular rotation and cause restrictive strabismus.




## Question:

The surgical photo right shows the lateral rectus muscle hooked and 2 small hooks under a white tissue above and below the lateral rectus muscle. Which of the tissues can be cut without causing the complication of fat adherence?

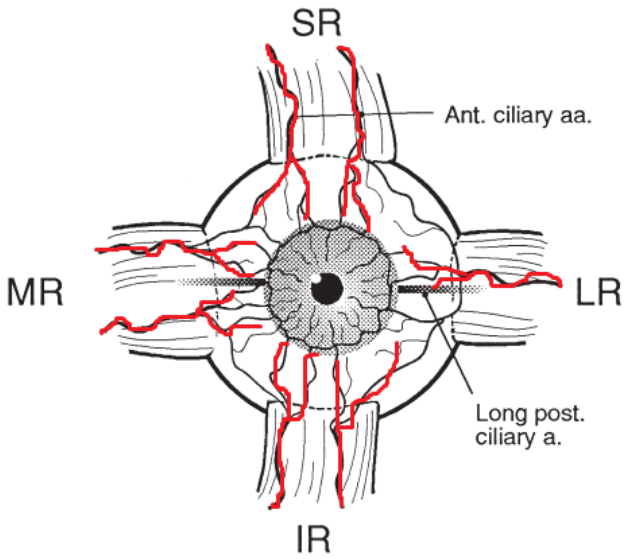



1. Tissue A
2. Tissue B
3. Both tissues A and B
4. None of the labeled tissues

**Answer:** In the surgical photo the tissue labeled "A" above and below the lateral rectus muscle is intermuscular septum and can carefully be cut without complication of fat adherence. It should not be incised more than 5 to 8 mm posterior in order to avoid cutting posterior Tenon's capsule. Tissue labeled "B" is the anterior part of the muscle sleeve (AKA pulley). This is where the lateral rectus muscle coursing posteriorly penetrates Tenon's capsule and becomes surounded by intra and extraconal fat. Violation of this tissue exposes orbital fat and can cause fat adherence to the muscle. Avoid surgical penetration of posterior Tenon's capsule and muscle sleeve. The answer is # 1.

## Anterior Segment Circulation

The two long posterior ciliary arteries and the anterior ciliary arteries supply circulation to the anterior segment. Each group contributes about 50% of the anterior segment blood flow (drawing right). The anterior ciliary arteries course with the rectus muscles. The MR, SR, and IR muscles having 2 arteries and are major suppliers, while the LR has one artery and contributes little to anterior segment circulation.

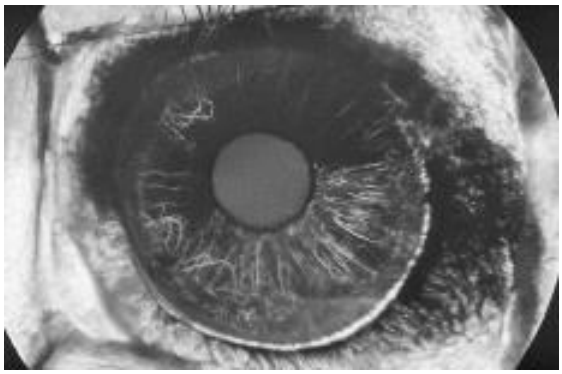



- **Major suppliers = MR, SR, and IR**
- **Minor contribution = LR**

Removing a rectus muscle will permanently disrupt the blood flow from the corresponding anterior ciliary arteries and the arteries do not re-canalize. In children the long posterior ciliary arteries can maintain enough flow, so even if all the rectus muscles were removed the child will not get **anterior segment ischemia**. In senior adults, however, the posterior ciliary supply can be compromised from small vessel disease and removing the 3 major supplier rectus muscles (i.e., MR, SR, and IR) can result in anterior segment ischemia. The rule of thumb is to preserve at least one major supplier to prevent anterior segment ischemia. Anterior segment ischemia can cause uveitis, hypotonia, and corneal edema. Anterior segment ischemia is usually transient lasting a few weeks to a couple months, however, severe cases can result in vision loss. Treatment is low dose topical corticosteroid drops. Anterior segment ischemia has been reported to occur 10 to 20 years after strabismus surgery.

## Question:

This is an iris angiogram taken after strabismus surgery that removed 3 rectus muscles. Which of the following describes the rectus muscle that was left intact.

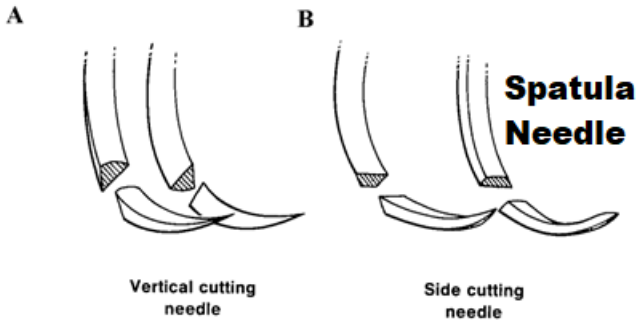


1. Elevator, adductor and intorter
2. Depressor, adductor, and extorter
3. Pure abductor
4. Pure adductor

**Answer:** The inferior iris is showing perfusion with hypo-perfusion above indicating an intact inferior rectus muscle with intact ciliary vessels. The inferior rectus muscle is a depressor, adductor and extorter so the answer is # 2. Note, once a rectus muscle is removed its ciliary vessels are permanently destroyed. In children collateral circulation develops, but in older adults anterior segment ischemia can be persistent. Because of the risk of anterior segment ischemia, it is recommended that strabismus surgery should leave at least one major vessel supplier rectus muscle (e.g., MR, SR, or IR) intact.

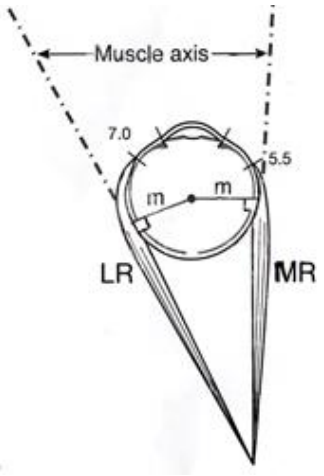
## Scleral Thickness

The scleral thickness behind the rectus muscle insertion is extremely thin, measuring only 0.3 mm. Because of the thin sclera, perforation into the globe is a significant risk during the scleral needle pass when suturing the muscle to sclera. The risk of perforation can be reduced by proper needle selection. The two basic types of needles are vertical cutting (Figure A below) and side cutting or spatula (Figure B below). The preferred needle for strabismus surgery is the **spatula (side cutting) needle** because it has a flat top and flat bottom. The flat bottom reduces the chance of inadvertent perforation deep into the globe and the flat top prevents cutting into the roof of the scleral tunnel above. The vertical cutting needle with the sharp pointed up or down can either penetrate too deep or cut through the roof of the scleral pass above.



# Chapter 2 Laws of Eye Movements

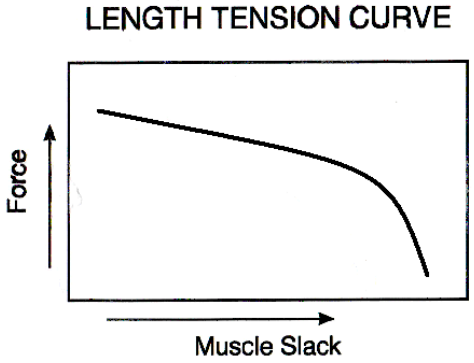
## Eye Movement




The force of eye rotation is proportionate to the length of the lever (moment arm "m") and the muscle force. The longer the lever (m) and the stronger the muscle force the greater the rotational force. The rotational force can be diminished by shortening the lever and by weakening the muscle.

**Rotational force = m X F**

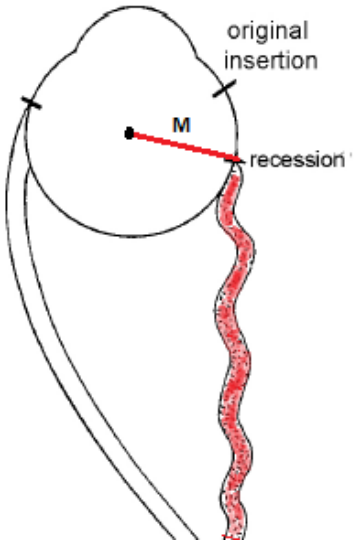
## Length Tension Curve

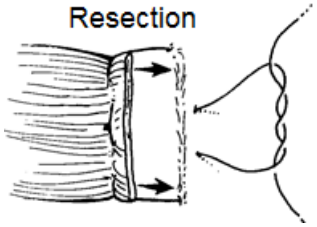



A muscle's ability to generate force is proportional to the tension or muscle slack. The more a muscle is slackened the weaker the muscle. This relationship is termed the length tension curve (Starling's Law). Note that the curve is not linear but exponential. Towards the end of the curve (to the right in the chart) a small increase in muscle slack causes a great reduction in muscle force.

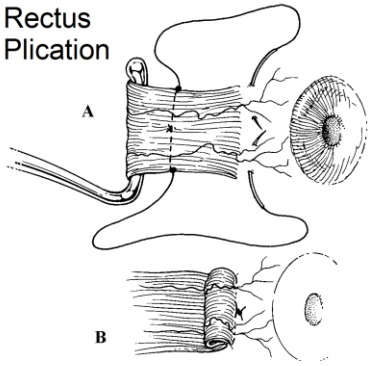
**Strabismus surgery**: There are two basic types of strabismus surgery: 1) ***recession*** that weakens a muscle by creating muscle slack, and 2) ***resection or plication*** that tightens the muscle. Reducing the moment arm, or changing the direction of muscle pull can also be used to correct strabismus.

**Rectus muscle recession:** The diagram right shows a rectus muscle recession. This is a weakening procedure that corrects strabismus by slackening the muscle to diminish muscle force. Because of the large arc of contact of the rectus muscles, a standard rectus muscle recession does not change the moment arm (see diagram right "m"). Thus, rectus muscle recessions work by creating muscle slack not by changing the moment arm. Weakening effect increases when the eye rotates to the recessed muscle as the slack increases.



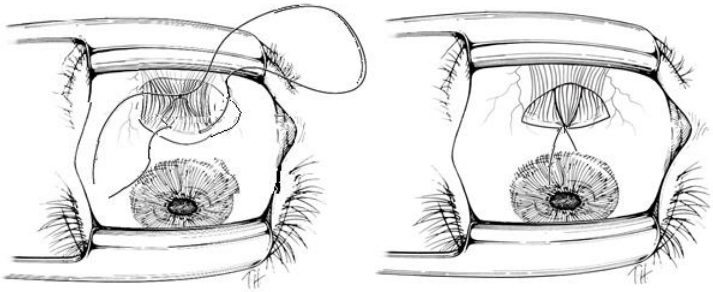



**Rectus muscle resection:** A rectus resection tightens the muscle by removing a segment of the muscle, then advancing the muscle to the original insertion. Tightening effect increases when the eye rotates away from the resected muscle because the muscle gets tighter).



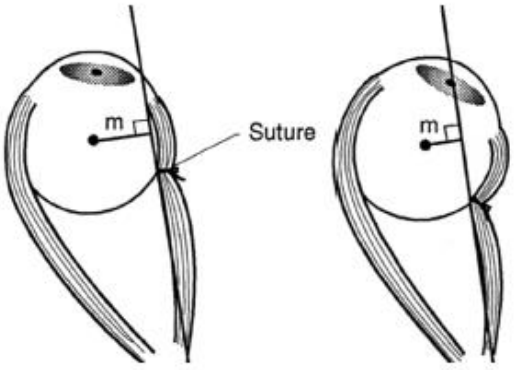

**Wright Rectus Muscle Plication** has the same effect as resection: it tightens the muscle. Sutures are attached to the muscle posterior to the insertion then passes through sclera anterior to the insertion (drawing right A). The sutures are pulled up to fold the muscle (drawing right B). Plication tightens the muscle without the need for muscle disinsertion so it is safer than resection. Plication spares the anterior ciliary vessels thus reducing the risk of anterior segment ischemia. The rectus plication was invented by your humble author (KW) during his fellowship and published in 1991. I like to call it the Wright plication but still waiting for that name to catch on – Darn!

**Wright Central Rectus muscle plication** is a modification of the full plication where only the center of the muscle is plicated (drawings right). Used for small deviations and corrects about 6 to 8 PD for each rectus muscle plicated.



**Faden Operation**

Faden is German for suture or twine and this procedure involves suturing the muscle to sclera behind the equator, approximately 14 mm posterior to the rectus muscle insertion. With the eye in primary position (center figure) the suture does not significantly alter arc of contact so the moment arm (m) is unchanged and there is no significant change in muscle slack. On rotation toward the faden muscle the muscle cannot release from sclera and the moment arm (m) is shortened (figure right). Thus the faden procedure only reduces rotational force when the eye rotates toward the faden muscle. Faden is used to weaken rotational force in the direction of the faden without having a significant effect in primary position. The faden is most effective when the fadened muscle is also recessed.

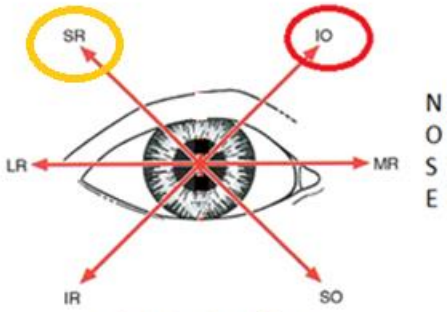


## Muscle action vs. Field of action

**Muscle action** is the direction the eye rotates away from primary position when a single extraocular muscle contracts. The muscle action is what is classically described in the tables in texts on anatomy and is summarized here:

| | Primary | Secondary | Tertiary |
|---|---|---|---|
| **SR –** | Elevation | intorsion | adduction |
| **IR –** | Depression | extorsion | adduction |
| **SO –** | Intorsion | abduction | depression |
| **IO –** | Extorsion | abduction | elevation |
| **LR –** | Abduction | | |
| **MR –** | Adduction | | |

**Field of action** is the position of gaze where an extraocular muscle is the primary mover of the eye. This position is where you move the eye to test an extraocular muscle's function. To test IO muscle function have the patient look up and nasal (red circle drawing right). To test SR muscle function the patient looks up and temporal (yellow circle drawing right). Field of action is the basis for the cardinal positions of gaze as shown in the drawing right.



Field of action

**Question:** The primary muscle action and field of action are in a similar direction for all of the following muscles except:

1. MR
2. IR
3. SR
4. SO

**Answer:** Primary action and field of action are in a similar direction for all the rectus muscles, but the oblique muscles are different. The SR muscle's primary action is elevation and field of action is up and temporal so they are similar - both up. The oblique muscle's primary action is torsion (IO extortion and SO intorsion) and field of action is up and in for IO, and down and in for the SO muscle so they are different. The correct answer is # 4, the SO muscle.

## Sherrington's Law of Reciprocal Innervation

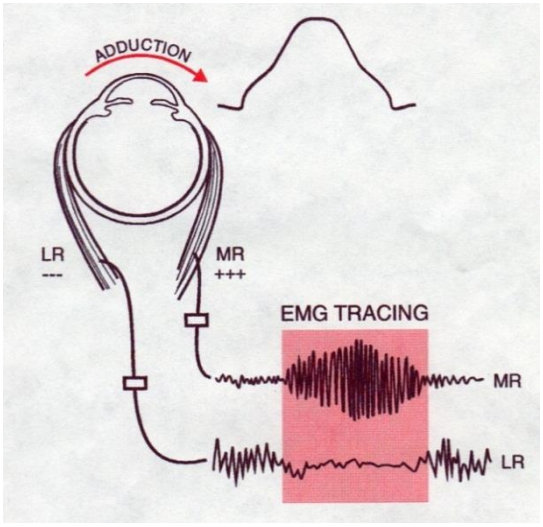
Sherrington's law states that when an eye moves there is an ***agonist*** muscle that contracts to move the eye and an opposing ***antagonist*** that relaxes allowing unopposed free rotation. For example on adduction the MR muscle contracts and the LR muscle relaxes. Sherrington's law relates to ductions, which is a single eye movement.

**Agonist – Antagonist Pairs**

- **M.R. --- L.R.**
- **S.R. --- I.R.**
- **S.O. --- I.O.**

Remember the "**S**" in **S**herrington is for **S**ingle eye!

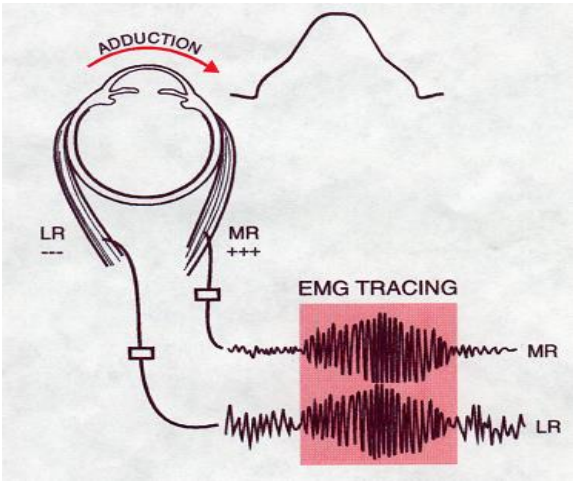
The diagram right shows physiology of Sherrington's law by an electromyogram (EMG). An electrode is attached to each of the horizontal recti. As the eye adducts, there is increased innervational activity of the MR muscle and inhibition of the LR muscle (red area). Note the baseline activity of the LR muscle is extinguished by inhibition during adduction.



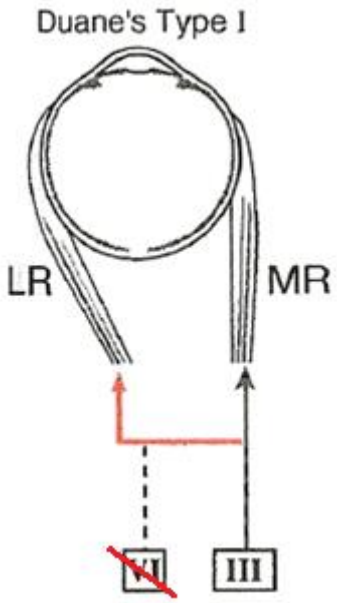

### Question:

What is the best explanation for the EMG right?

1. Normal physiology of Sherrington's law
2. A violation of Sherrington's law caused by aberrant innervation with LR inappropriately contracting on adduction.
3. The MR and LR show signs of neuromuscular disease
4. Violation of Listing's law of ocular rotation.

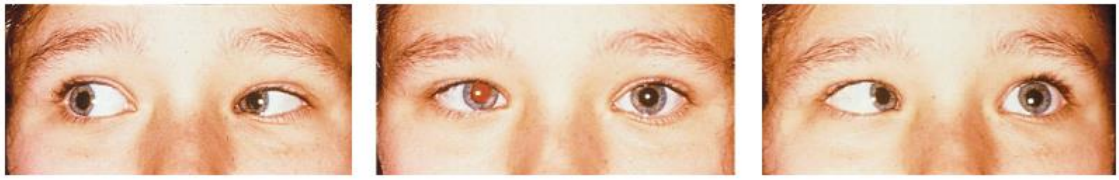


## Answer:




This EMG shows aberrant innervation with the MR and LR muscles co-contracting at the same time as the eye adducts (see red area in diagram in the question). The LR muscle is inappropriately contracting on adduction- it should be inhibited. This is a violation of Sherrington's law. The answer is # 2. The EMG in this question is typical of Duane's syndrome, which is congenital agenesis of the 6th nerve and nucleus with misdirection of part of the MR nerve (3rd nerve) to innervate the LR muscle (see drawing right). As the patient adducts the MR branch of the third nerve stimulates both MR and LR muscles causing co-contraction.

Type 1 and 3 Duane's syndrome exhibit limited abduction (sixth nerve palsy), globe retraction and lid fissure narrowing on adduction. Note in photos below is a left Duane's syndrome showing limited abduction left eye with lid fissure widening and left lid fissure narrowing on adduction. Globe retraction and lid fissure narrowing on adduction is caused by the co-contraction of MR and LR muscles that pulls the eye back posteriorly.

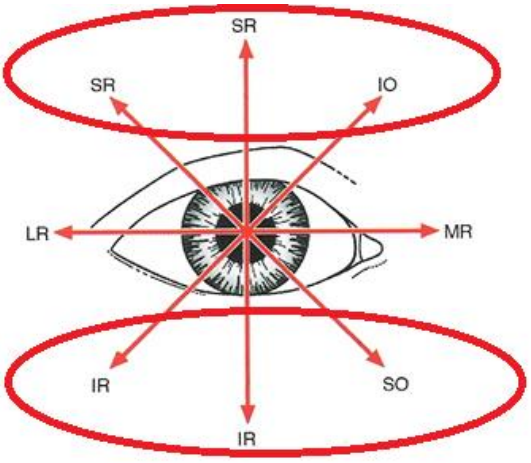


**Duane's syndrome left eye:** See left eye has limited abduction and on adduction there is lid fissure narrowing.

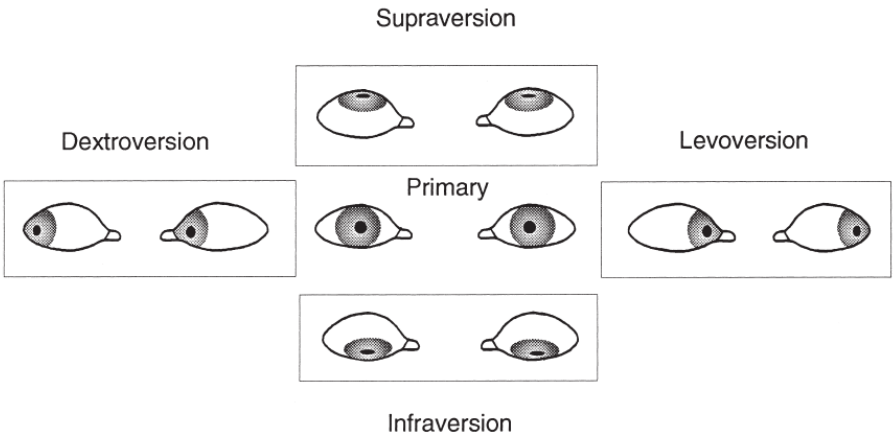
## Synergists

Synergist muscles are simply muscles that move the eye in the same direction. For example:

- **Supraduction** - ***S.R. & I.O.*** (Red oval above)
- **Infraduction** - ***I.R. & S.O.*** (Red oval below)



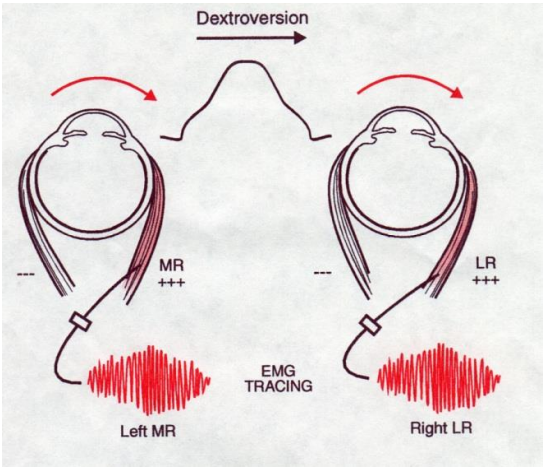
## Versions




Versions are binocular movements. They are used to determine how well the eyes move together. Mild muscle palsy or restriction could be missed on duction testing, but is seen on version testing where you can compare eye movements. Oblique muscle overaction is also seen on versions but not seen on ductions.

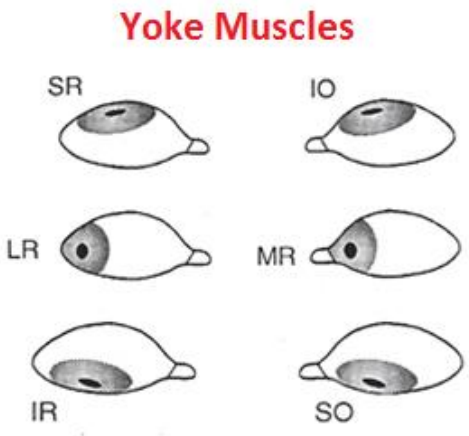
## Hering's Law of Yoke Muscles

Hering's law states that versions occur as "yoke" muscles from both eyes receive equal innervation so the eyes move together - can't move just one eye. Yoke muscles are agonists and antagonists that move the eyes in the same direction. For example; on dextroversion the left MR and right LR are yoke agonists muscles. The left LR and right MR are yoke antagonist muscles. Diagram right shows dextroversion as the yoke agonist muscles left MR muscle and right LR contract (red) while the antagonist yokes left LR and right MR muscles relax.



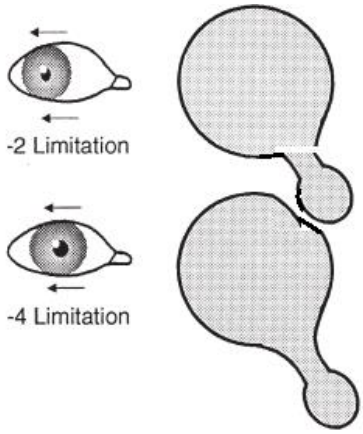

Remember: The **H** in Hering has two vertical lines linked together by a horizontal bar like the two eyes that are yoked together.

*Ok! I agree this "H" thing maybe kind of lame, but that is the best I could come up with!*



## Quantitation of Ductions

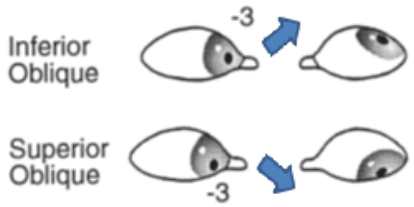
Limitation of ductions can be quantitated on a scale of 0 to -4. Minus 1 is minimal limitation and -4 is severe limitation of ductions with the eye not moving past midline (see drawing right). This scale is a rough subjective estimation.



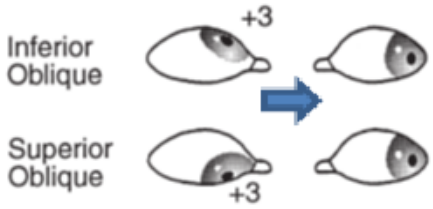

## Quantitation of Versions

Abnormal versions can be quantified as overaction or under action on a scale of + 4 severe overaction, to - 4 severe under action. Oblique overaction is only seen on version testing.

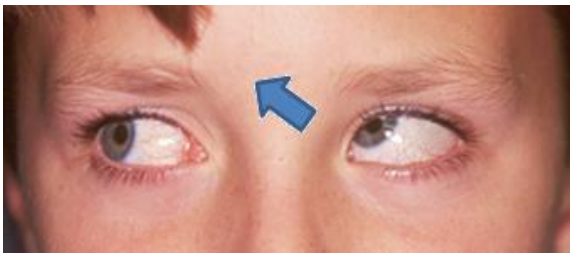
***Oblique Under action***




***Oblique Overaction***




**Question:** Which muscle or muscles may be dysfunctional in this patient looking up and right?



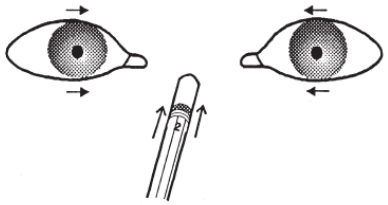
1. Left IO overaction
2. Left SO under action
3. Right SR weakness
4. Right IR restriction
5. All of the above

**Answer:** The photo shows abnormal versions with an up-shoot of the left eye in right gaze. At first look this appears to be a left IO muscle overaction, and yes this is a possibility. Per Sherrington's law of agonist and antagonist muscles the apparent IO overaction could also be due to a left SO under action. Another possibility is the left eye up-shoot could be secondary to limited elevation of the right eye in right gaze. If the patient fixes with the right eye and it has difficulty elevating the eye in abduction, this would cause a secondary overaction of the yoke left IO muscle. As the right SR muscle works hard to move up and out the left IO (yoke of the SR) also gets overstimulated and overshoots. Remember: per Hering's law yoke muscles get equal innervation. Causes of limited elevation in abduction are either a weak SR muscle (field of action is up and out), or a tight IR muscle. The answer is # 5, all of the above.

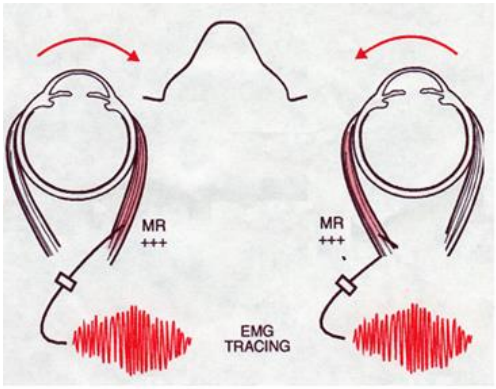
## Motor Fusion - Vergence Eye Movements

Motor fusion is a mechanism that keeps our eyes locked on target. Motor fusion invokes correctional movements called vergence movements that keep our eyes from drifting off target and for following an approaching target or a target moving away. There are three vergence movements: convergence, divergence, and vertical vergence.

When a target approaches we use convergence to bring the eyes together on the target (drawing right). When a target goes away we use divergence to separate the eyes keeping alignment on the target. Vertical vergence keeps our eyes from drifting vertically off target.



The diagram right shows an EMG tracing of both MR muscles contracting (red) at the same time in the act of convergence. Note that vergence violates Hering's law of yoke muscles used for versions. You can demonstrate convergence by fixating on the tip of a pencil then bringing it in to your nose, and divergence by moving the pencil away. Note our convergence is much stronger than divergence and vertical vergence is almost nonexistent.



Convergence

### *Vergence Fusional Amplitudes*

Convergence 25 PD, Divergence 8 PD, and Vertical 1-2 PD

**Question:** Which of the following are true about convergence?

1. Corrects for our innate tendency for the eyes to diverge
2. Abides by Sherrington's Law
3. Violates Hering's Law
4. Strongest vergence amplitudes
5. All of the above

**Answer:** Convergence corrects our natural divergence so # 1 is correct. During convergence the medial rectus muscle contracts and the lateral rectus muscle relaxes consistent with Sherrington's law of agonist-antagonist (S for single eye) so # 2 is correct. Convergence involves contracture of MR muscles of each eye simultaneously, thus violating Hering's law of yoke muscles so # 3 is correct. Convergence is our strongest version so # 4 is correct. The answer is # 5, all of the above are true.

## Saccadic Eye Movements

- First eye movements occurring 1-2 weeks after birth
- Fast 250 deg/sec
- Ballistic - preprogrammed contraction with no visual feedback
- Foveal vision suppressed
- Need good muscle function

Saccadic eye movements are fast dart like primitive movements. They are our first eye movements and are why newborns have jerky eye movements. In contrast to smooth pursuit eye movements, saccadic eye movements are not controlled by visual feedback as there is foveal suppression during a saccade. A saccade is ballistic like a football thrown by a quarterback. After the ball leaves the quarterback's hand the trajectory cannot be changed. The job of a saccade is to move the eyes fast and close to the target then smooth pursuit accurately zeros in on the target. Approximately 70 to 80% of muscle strength is required to generate a saccade so the presence of a saccadic eye movement indicates good muscle function. Inability to perform saccade is a sign of extraocular muscle paresis.

You can experience saccadic foveal suppression by looking as fast as you can right – left or up –down. Note your vision extinguishes during the saccade.

**Question:** A patient has an eye with limited abduction. The eye moves rapidly from adduction temporaly then abruptly stops at the midline. The limited abduction is caused by lateral rectus muscle palsy. True or False?

**Answer:** The ability to generate a saccade indicates good muscle strength, so the fast abduction movement indicates good lateral rectus muscle function not muscle palsy, so the answer is false. The reason for the limited abduction is restriction, not a muscle palsy. Restricted abduction can be due to a tight medial rectus muscle or a scar to the eye ball that restricts rotation.

## Smooth Pursuit Eye Movements

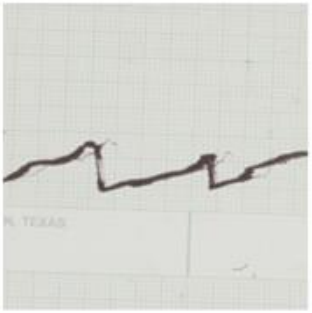
- Slow accurate eye movements to follow a target < 30 deg/sec
- Requires foveal fixation
- Develop after 2 – 3 months of age

Smooth pursuit eye movements are slow accurate movements used to fixate and follow a target. Smooth pursuit develops after saccadic movements at around 2 to 3 months of age. They require foveal fixation and visual feedback to keep the eyes on target. In contrast to a saccade which is a ballistic unguided movement, smooth pursuit movements are analogous to a laser guided missile and locks on the target. Poor fixation and imprecise following of a visual target can indicate poor vision.

## OKN Optokinetic Nystagmus

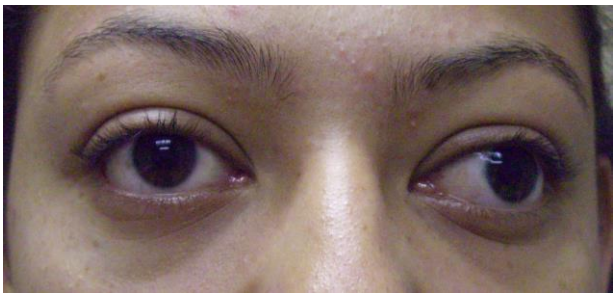
This is the reflex of the eyes following a slow moving target with smooth pursuit until the end of excursion then re-fixation with a saccade as the next target comes into view. OKN drum spinning slowly will stimulate smooth pursuit in one direction, then a saccade in the opposite direction to re-fixate on the next target. The eye movement recording below shows normal saw tooth pattern of OKN stimulation. The acute vertical up and down line tracings are the fast saccades and the gradual up horizontal tracings are the slow smooth pursuit movements.

OKN Drum



Normal OKN Tracing

**Question:** Patient in photo had left medial rectus surgery and developed a large an exotropia (left eye turns out) immediately after surgery. Left eye shows limited adduction, the eye adducts slowly, and there is no adduction saccade.



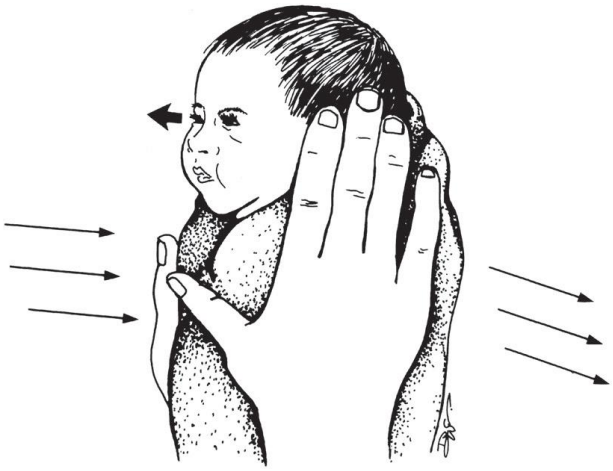
Which is most likely?

1. Rectus muscle is weak caused by the surgical complication of a lost muscle.
2. Left lateral rectus muscle is tight causing restriction and the medial rectus function is good.
3. Surgical complication of scarring and restriction
4. # 2 and #3 are correct

**Answer:** Left eye has slow adduction and no adduction saccade indicating a weak medial rectus muscle. The weak medial rectus is responsible for the exotropia. It is likely that there was the complication of a lost medial rectus muscle at the time of surgery so the answer is #1. The medial rectus muscle is the muscle most commonly lost during strabismus surgery as it has no connection to an oblique muscle. The limited adduction is secondary to a weak medial rectus muscle not restriction so answers B and C are wrong.

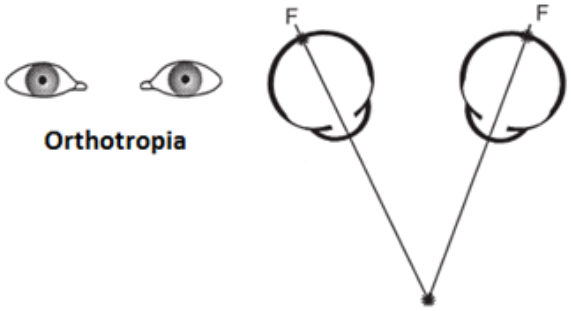
## Dolls Head Maneuver

If an infant or child is uncooperative you can examine ocular rotations by the dolls head maneuver or by spinning the child. Move the head to one direction and the eyes will move to the opposite direction. This movement is based on vestibular stimulation. See drawing right that shows the baby being moved to the baby's left and the eyes go to the baby's right. If there was a right 6$^{th}$ nerve palsy the right would show limited abduction.



# Chapter 3 Introduction to Strabismus

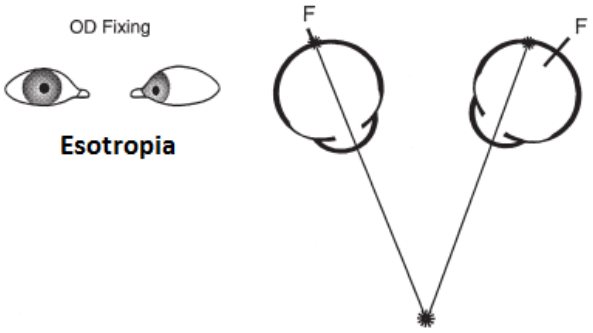
Normally the eyes are aligned so both foveas (F) are on target, and this is termed **orthotropia** (drawing upper right).




**Strabismus** is defined as eye misalignment or a tendency for an eye to drift so one fovea is off target. Strabismus can be horizontal, vertical, torsional, or a combination of the three.

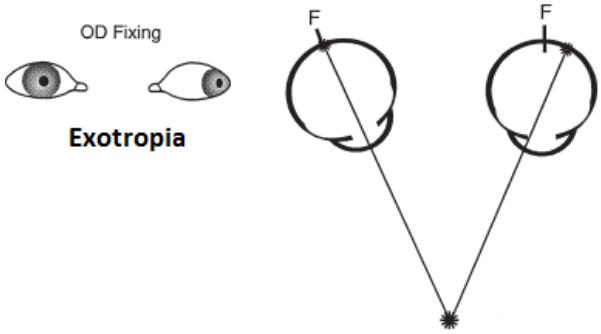
## Tropia

***Tropia*** means the eyes are misaligned with one fovea on target (fixing eye) and the other fovea off target (deviated eye).



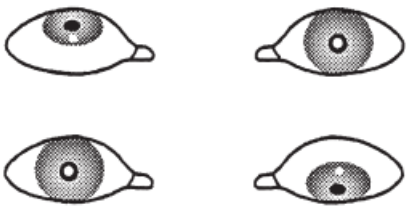

***Esotropia (ET)*** - the deviated eye turns in so the image falls on nasal retina (drawings right).

***Exotropia (XT)*** - the deviated eye turns out so the image falls on temporal retina (drawings right).

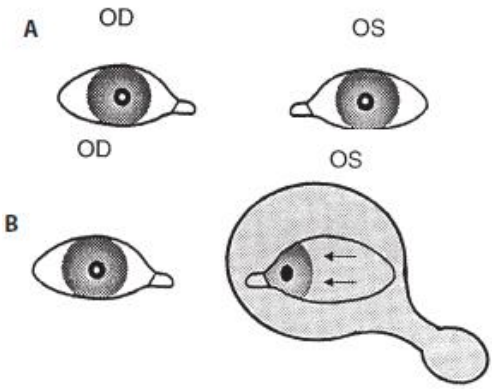



***Cyclotropia*** means the eye is twisted: ***extorsion*** 12 o'clock temporal and ***intorsion*** 12 o'clock nasal.

***Hypertropia (HT)*** - the deviated eye turns up so the image falls on superior retina. ***Hypotropia*** the eye turns down so the image falls on inferior retina. Note with the left eye fixing (upper drawing) there is a right hypertropia (RHT) and when the right eye fixates there is a left hypotropia. This is the same deviation. By convention we usually use the hypertropia (HT) term to describe vertical tropias.



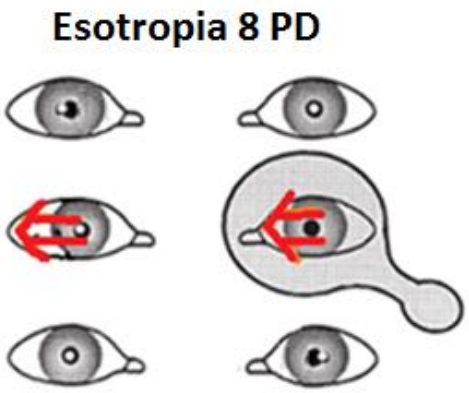
## Phoria




While a tropia is a manifest deviation, a phoria is a latent deviation held in check by binocular fusion. It is a tendency for an eye to drift off target but binocular fusional vergence holds the eyes in alignment. The diagram right depicts an esophoria. Figure A shows eyes aligned, no deviation as the patient is fusing. Figure B shows covering the left eye to break binocular fusion. Once fusion is suspended the occluded eye turns in and the latent esophoria becomes manifest. If the fusion is strong the eye must be occluded for several seconds to disrupt fusion as a quick cover-uncover will not manifest the phoria. When the occluder is removed the patient will diverge and regain alignment and binocular fusion. Virtually every normal person has a small phoria that is held in check with binocular fusion. Exophoria is most common because our orbits diverge 23 degrees, and convergence keeps alignment. A phoria may spontaneously become manifest and turn into a tropia if the patient is tired, run down by illness or takes a sedative like valium or alcohol. *Be careful, alcohol can bring out deviant tendencies or so I have been told*. An intermittent tropia is noted by parenthesis around the "T" so an intermittent exotropia is written X(T).

## Cover-Uncover Test (Tropia Test)

Cover-uncover test is used to detect a small tropia without disrupting binocular fusion because we don't want to bring out a phoria. We only want to know if there is a tropia. Cover-uncover testing is not needed for large tropias as the tropia will be obvious.




*Performing Cover-Uncover Test:* Briefly cover then uncover the eye that is presumed to be fixing. If there is no re-fixation movement, then cover-uncover the other eye. No re-fixation shift from either eye indicates orthotropia. Drawing right shows a micro right ET. Note that covering the fixing left eye causes a re-fixation and both eyes move right so the right eye comes to fixate. Both eyes move to the right because of Hering's law. Bottom diagram now shows the right eye fixing and a small left esotropia.

Note: Brief cover-uncover for one to two seconds does not disrupt fusion and prevents the manifestation of a phoria. Cover too long (> 3 seconds) and you will break fusion and manifest a latent phoria.

## Alternate Cover test (Phoria Test)

Alternate cover test suspends fusion so the full deviation is measured - both phoria and tropia. Binocular fusion is disrupted by covering one eye for several seconds then quickly moving the cover to the fellow eye so the patient does not have time to regain fusion. The alternate cover test keeps binocular fusion suspended as one eye is always covered.

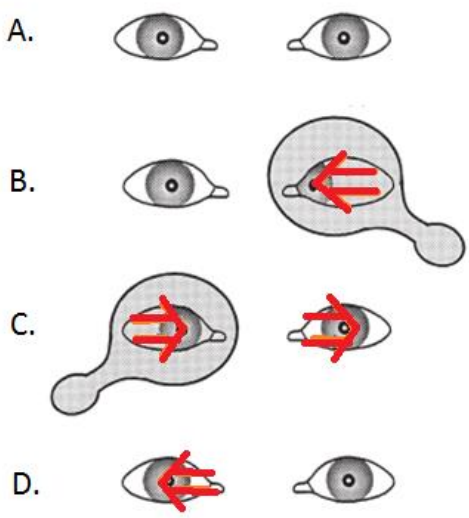
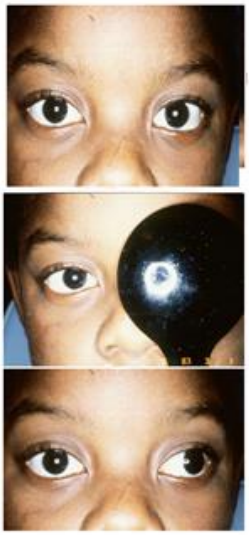



Diagram right shows an esophoria. Drawing "A" shows orthotropia patient fusing. "B" shows left eye covered for several seconds and the eye under the cover drifts in because fusion is broken. "C" shows the cover quickly moved over the right eye which causes the left eye to move to the left to pick up fixation. Note the right eye under the cover also moves left because of Hering's law. "D" shows the cover removed and the patient regains fusion as the right eye diverges to restore alignment.

### Question:



The patient right shows covering left eye for 10 seconds then removing the cover. This patient is showing -

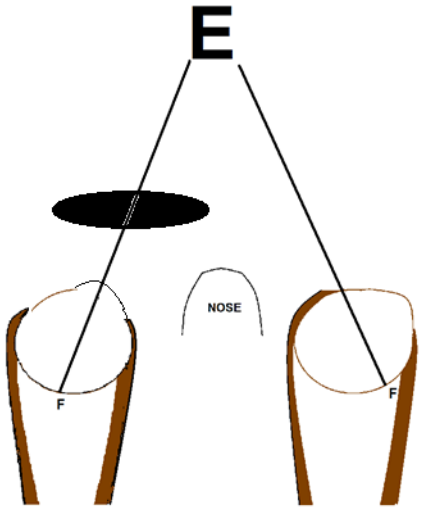
1. Orthotropia
2. Exophoria
3. Convergence
4. Binocular fusion
5. All of the above

**Answer:** This is a photo series of a patient with a large exophoria. Top photo shows orthotropia as Hirschberg light reflexes are centered each eye. Right middle photo shows covering left eye which breaks fusion. The cover is removed (lower photo) disclosing that there is a large exophoria which had been controlled by fusional convergence. The ability to align the eyes by converge indicates binocular fusion is present. The correct answer is # 5 as all are correct. If the exodeviation spontaneously becomes manifest the strabismus is considered an intermittent exotropia; however, the term exophoria also applies.

Note: the term ***"dissociate"*** the eyes means to break binocular fusion.

## Positive Angle Kappa

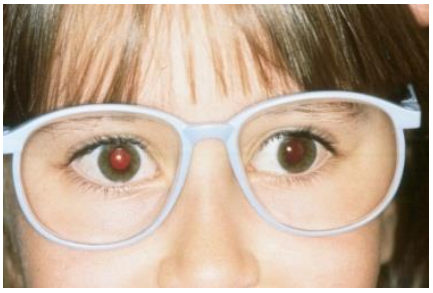
Line of sight is the line between the visual target and the fovea. If the fovea is displaced temporally in the newborn infant the eye will adapt by turning out so the fovea aligns with the visual target. The eye will look exotropic, but on cover testing there is no re-fixation movement as the foveas are aligned on the target. Diagram right shows a positive angle Kappa with the right fovea displaced temporally. The right eye is oriented in a divergent position so the fovea can align with the visual target (**E**). Note that even when the left eye is covered the right eye stays divergent as this is the position where the fovea is aligned on the target. Prematurity associated severe ROP and temporal dragging of the fovea is a common cause of a positive angle Kappa.

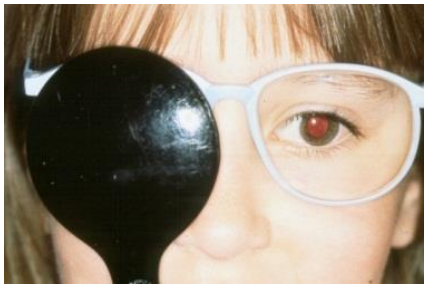



***Negative angle kappa*** is when the fovea is displaced nasally the eye turns in to align with the target. Negative angle Kappa is rare. The eye looks esodeviated but there is no re-fixation shift on cover testing.

***Note:*** Strabismus goes away when you cover one eye (as long as ductions are full). Angle Kappa is not strabismus as it does not go away when an eye is covered. It is an ectopic fovea from a retinal problem.

**Question:** Photos right – The most likely diagnosis is:



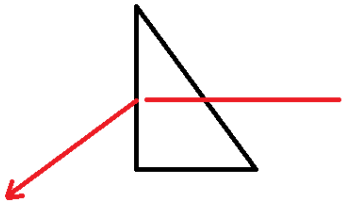


1. Exotropia
2. Exophoria
3. Intermittent exotropia
4. Orthotropia history of ROP

**Answer:** There appears to be a left exotropia in upper photo. Lower photo shows right eye covered, yet the left eye stays in a divergent position. A monocular divergent eye indicates a positive angle Kappa, not an exotropia. This patient has a history of prematurity and ROP. Fundus exam shows temporal dragging of the fovea left eye. The best answer is # 4 as the patient is orthotropic with a positive angle Kappa.

## Prisms

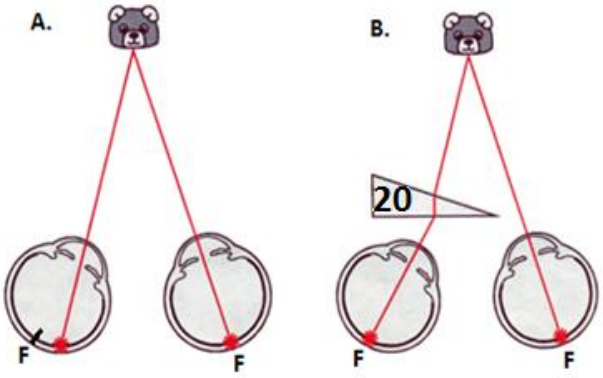
Prisms bend light to the base of the prism. The deflection of light is measured in prism diopters (PD) where one prism diopter equals about ½ a degree.



## Prism Neutralization

Prisms can be used to neutralize or correct strabismus by directing the images so they land on both foveas. The prism that neutralizes the deviation is the measurement of the deviation in prism diopters (PD). Prism neutralization can be done by placing the prism over the deviated eye or the fixing eye.

**Prism over deviated eye**: Placing prism over the deviated eye neutralizes the strabismus by deflecting the eccentric image onto the fovea. Figure A (right) shows an esotropia with the left eye turned in and the image falling nasal to the fovea (F). The right eye is fixing on the target so the image is on the fovea. In figure B, a 20 PD base out prism placed over the deviated left eye to deflect the image onto the fovea thus correcting or neutralizing the esotropia. This is a 20 PD esotropia.

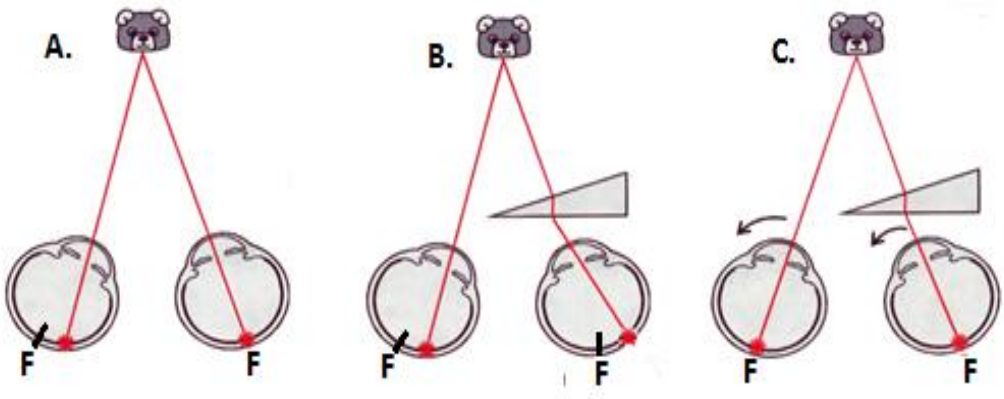



To neutralize a deviation orient the prism so the apex points in the direction of the deviated eye as seen in figure B above:

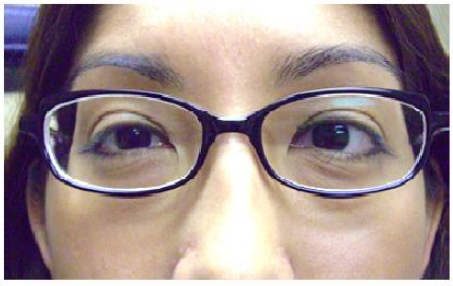
- ET apex points nasally and base out (BO)
- XT apex points temporally and base in (BI)
- Hypertropia apex points up and base down (BD)
- Hypotropia apex points down and base up (BU)

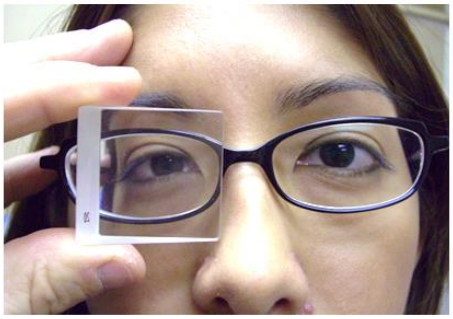
**Prism Over Fixing Eye:**

One can prism neutralize a strabismus by placing the prism over the fixing eye to shift the eyes into position using Hering's Law. Drawing A (below) shows a left esotropia with right eye fixing. Figure B shows the prism placed BO over the fixing right eye which deflects the image off the fovea onto temporal retina. The patient sees the image has moved. Figure C shows the patient re-fixating on the target to place the image back on the fovea. The fixing eye initiates the re-fixation, moving nasally and the non-fixing eye simultaneously moves in tandem temporally because of Hering's law of yoke muscles. Note in figure C both eyes move to the left (levoversion) and now both retinal images are on the foveas so the deviation is neutralized. Placing a prism over the fixing eye will cause both eyes to move in the direction of the apex.




**Question:** Photo top right shows a patient with an esotropia fixing right eye because the left eye is blind. Bottom photo shows base out prism over the fixing right eye.





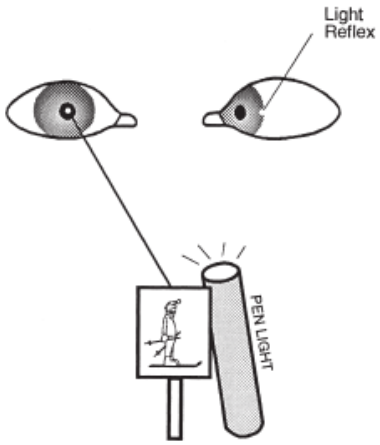
Which of the following is true regarding this patient?

1. Left eye is fixing
2. BO prism over right eye moves both eyes left per Hering's law
3. BO prism induces convergence
4. It is wrong to place the prism over fixing right eye for prism neutralization

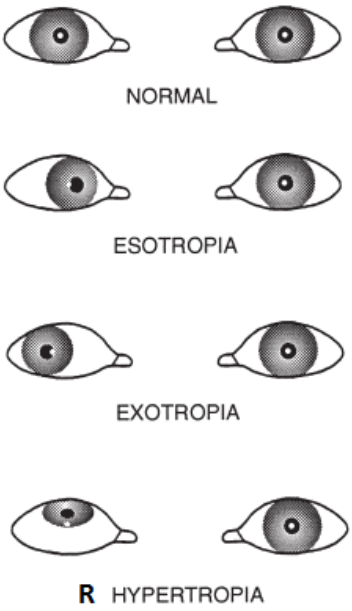
**Answer:** Top photo shows right eye fixing and the blind left eye eso**tropic**, so # 1 is wrong. In the lower photograph a base out prism is placed over fixing right eye which induces a version movement to the left. This is Hering's law that produces levoversion as both eyes move to the left when the prism is placed over the fixing right eye so answer # 2, Hering's law, is correct. This is a version movement, it is not convergence movement so # 3 is wrong. Placing the prism over the good eye (i.e., fixing eye) is an excellent method for measuring strabismus associated with a blind eye (sensory strabismus), so # 4 is wrong. Measure the deviation by increasing the prism over the fixing eye until the deviated eye comes to midline.

## Hirschberg Test

Alignment can be estimated by shining a light into the patient's eyes to create a light reflex in the pupils (drawing right). The reflex is sometimes called the corneal light reflex, but it is actually the first Purkinje image which is a virtual image of the light located a couple millimeters behind the pupil. The Hirschberg Test is only done at near. The Hirschberg test cannot be done at the distance as it is not possible for the examiner to see the light reflex location in the pupils when the examiner holds the light in the distance (20 feet). It is best to have the patient look at an accommodative target placed next to the light as shown in the figure right.



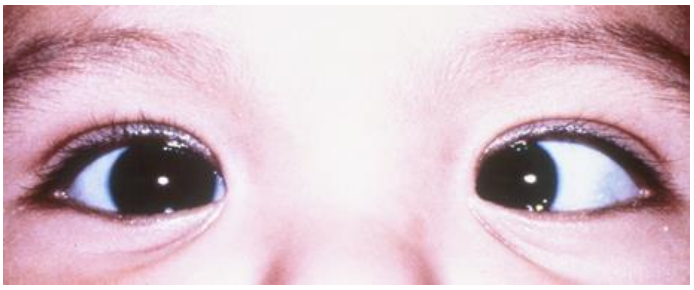

If the reflexes are centered in the pupil to a slight symmetrical nasal deviation (< 5 degrees) the eyes are in normal alignment called orthotropia (see drawing right). The slight nasal displacement is normal and called physiologic positive angle kappa. Temporal deviation of the reflex means esotropia, nasal deviation of reflex indicates exotropia, and vertical displacement means hypertropia.



**Estimate of the deviation by reflex displacement:**
One can make a rough estimate of the size of the deviation by observing the location of the light reflex in the deviated eye.

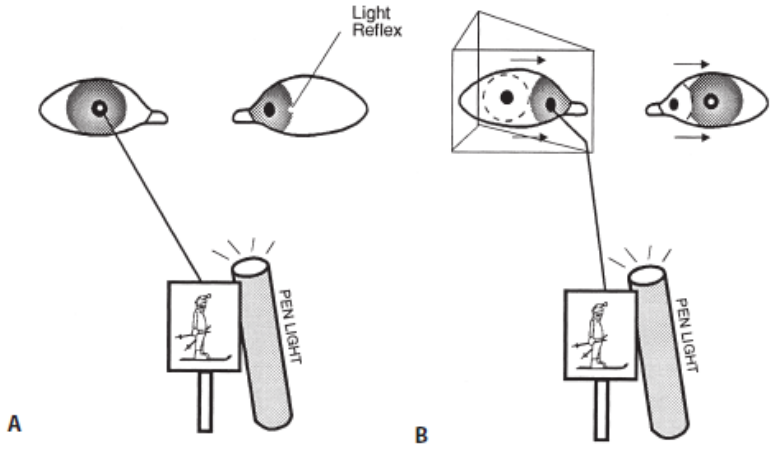
- Pupillary margin 30 PD
- Mid iris 60 PD
- Limbus 80 PD

Photo right shows the light reflex left eye is at mid iris so the deviation is estimated at ET 60 PD.



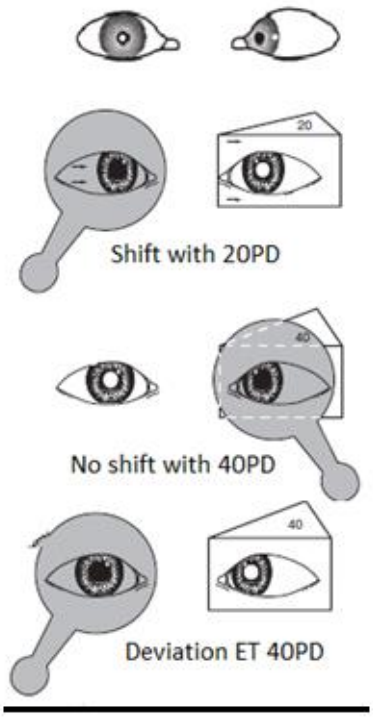
## Krimsky Light Reflex Test

The Krimsky test uses the first Purkinje image reflex like the Hirschberg test but adds prism neutralization to measure strabismus. With the Krimsky test a prism is placed over either the fixing eye or the deviated to center the reflex in the pupils. The figure below shows the Krimsky test placing the prism over the fixing eye. Figure A shows a left esotropia with fixation right eye. A light is placed next to a fixation target and is directed to both eyes. Next as shown in figure B a prism is placed in front of the fixing right eye which moves both eyes by Hering's law to center the light reflex. Various prisms are used until the light reflex is centered. This is similar to neutralizing the deviation by placing the prism over the fixing eye as described in detail above and shown in the question on page 36.



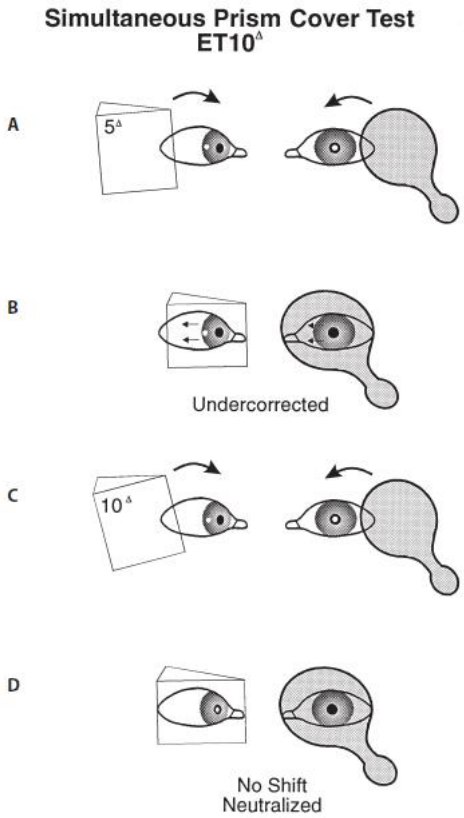
## Prism Alternate Cover Test

Prism alternate cover test is used to break up fusion in order to measure the full deviation: phoria and tropia. One eye is always occluded so fusion is suspended thus the phoria will become manifest. Diagram right shows an esotropia. Place a prism over the deviated eye and alternate the cover to see if there is a refixation shift. Increase the prism until there is no shift to alternate covering of the eyes and this prism is the measure of the deviation. This is usually the measure that provides the basis for the amount of strabismus surgery.




## Simultaneous Prism Cover Test

This test is like the cover un-cover test because its purpose is to examine the tropia only, without manifesting a phoria. The difference is the simultaneous prism cover test actually measures size of the tropia, whereas the cover-uncover test just identifies that a tropia is present. The key is to only briefly cover the fixing eye and not to break fusion. Diagram right shows (A) left fixing eye covered simultaneously with a 5 PD prism BO placed over the deviated right eye. Figure (B) shows 5 PD was not enough as right eye refixates. Figure (C) shows increasing the prism to 10 PD BO and again prism and cover are simultaneously placed with prism over the deviated eye, and occluder over fixing eye. Note they are left in place for only one to two seconds then removed so to not break fusion. Figure (D) shows there is no re-fixation movement with 10 PD BO so the tropia measures esotropia 10 PD. Once the tropia is measured by simultaneous prism cover test then the full deviation phoria and tropia can be measured with prism alternate cover test as described above. Note: The diagram exaggerates the appearance of the esodeviation for demonstration purposes.



**Question:** If simultaneous prism cover test measures esodeviation 5 PD and prism alternate cover test measures esodeviation 20 PD the full esodeviation is 25 PD. True or False?

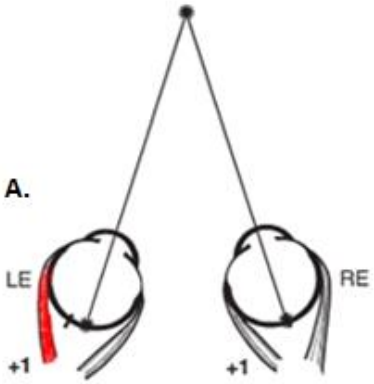
**Answer:** The prism alternate cover test measures the full deviation tropia and phoria together so the full deviation measures 20 PD not 25 so the answer is false. Simultaneous prism cover test measures only the tropia so the tropia is 5 PD. The phoria is 15 PD (full deviation 20 PD minus the tropia 5 PD). Operate for the fully dissociated deviation of 20 PD.

## Primary vs. Secondary Deviation

Strabismus associated with limited eye movement can be caused by either restriction of ocular rotation or extraocular muscle paresis. The primary deviation is when the "good eye" fixes on the target, and the secondary deviation when the eye with limited movement is forced to primary position to fixate. The secondary deviation is larger than the primary deviation. The secondary deviation is greater because it takes great effort for the eye with limited ductions to get to primary position. Because of Hering's law both eyes receive increased innervation so the good eye will greatly overshoot (see figures A and B).

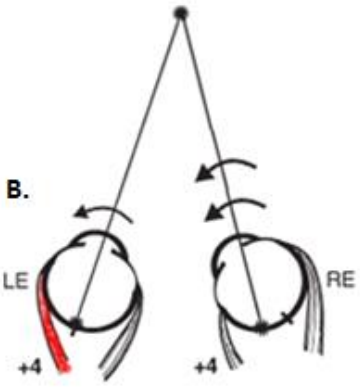
Figures A and B (right) show an esotropia caused by a paretic left lateral rectus muscle (red shading).

Figure A (right) shows the primary deviation with the right "good eye" fixing, and a 25 PD esotropia. Minimal innervation is needed to hold the good eye on target in primary position. The left eye is turned a little because of the weak left lateral rectus muscle.




ET 25 PD
Primary Deviation

Figure B (right) shows the secondary deviation with the paretic left eye forced into primary position in order to fixate on the target. Since the left LR muscle is weak extra innervation is required to abduct the left eye into primary position. Because of Hering's law the yoke right MR muscle also receives extra innervation and since it is strong the right eye turns way in (double arrows) causing a big secondary esotropia (ET 50 PD).

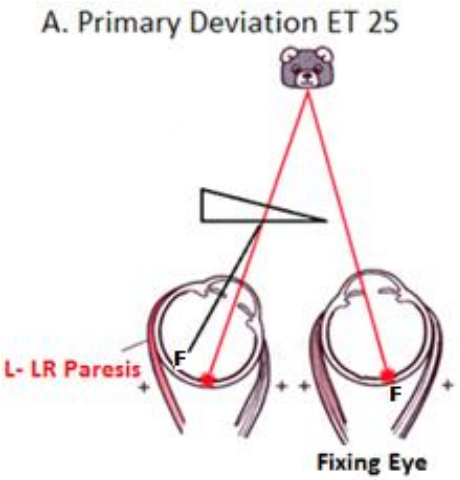



ET 50 PD
Secondary Deviation

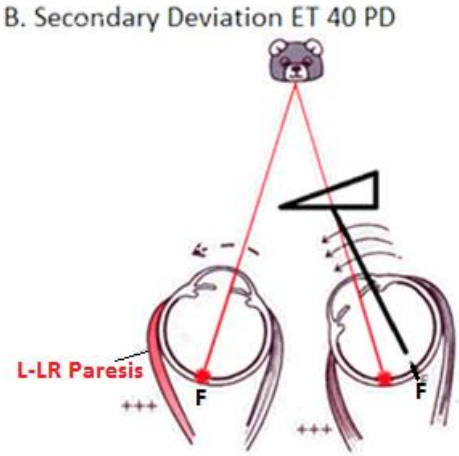
## Measuring Primary vs. Secondary Deviation

Figures A and B show a left lateral rectus muscle paresis causing a left esotropia with the good right eye fixing.

**Primary Deviation:** Figure A (right) shows measuring the primary deviation by placing a base out prism over the paretic left eye. Since the prism is over the paretic eye, the paretic eye can stay deviated in its position of rest so all muscles receive minimal innervational activity and the esotropia is small. *Measure the primary deviation by placing the prism over the paretic eye.*




**Secondary Deviation:** Figure B shows measuring the secondary deviation by placing a base out prism over the "good" eye to force the paretic eye to abduct and move into primary position per Hering's law. Note the right eye must turn way in in order force the weak left lateral rectus muscle to abduct and bring the left eye to fixate on the target. A large prism must be placed over the "good eye" to force the paretic left eye into primary position. The secondary deviation is larger than the primary deviation. *Measure the secondary deviation by placing the prism over the "good eye".*

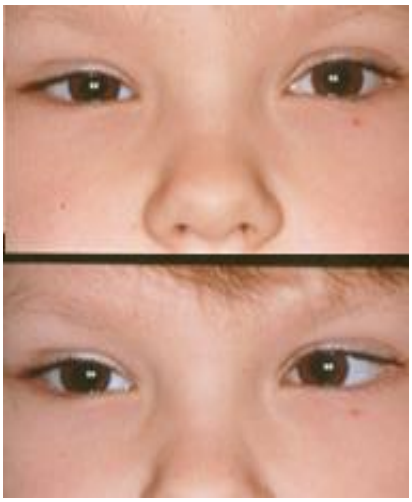



### *Remember*

- ***Primary deviation (Small)*** **-** When the good eye is fixing and neutralizing prism is over paretic eye.

- ***Secondary deviation (Large)*** **-** When the eye with the paretic muscle is forced to fixate in primary position. Neutralize with prism over good eye.

**Question:** This 5 year old in photos right has an esotropia secondary to a postviral 6th nerve palsy. Note that the esotropia is greater when right eye is fixing (lower photo). All are true except:



1. Right 6th nerve palsy
2. Neutralize deviation with base out prism
3. Primary deviation is neutralized with prism over right eye
4. Secondary deviation is neutralized with prism over left eye
5. Limited abduction left eye

**Answer:** First determine which eye has the 6th nerve palsy. The deviation is greater with the paretic eye fixing. Since the bottom photo with right eye fixing shows the larger deviation there is a right 6th nerve palsy. Yes you neutralize an esotropia with a BO prism, apex pointed in. Neutralize the primary deviation by placing the prism over the paretic right eye, so it stays in the position of rest. Likewise the secondary deviation is neutralized by placing a prism over good left eye, which forces the paretic eye to fight come to primary position. The answer is # 5, limited abduction left eye – as the left eye is the normal eye.

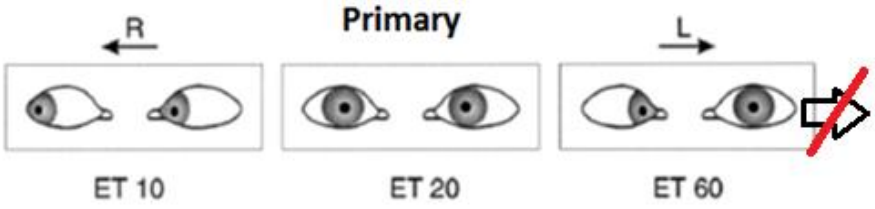
## Comitant vs. Incomitant Strabismus

If a strabismus is the same in all fields of gaze the strabismus is termed ***comitant*** as shown below.

| | Right Gaze | Primary | Left Gaze |
|---|---|---|---|
| **Comitant -** | ET 20 | ET 20 | ET 20 |

If the ductions are limited then the deviation will be different in different fields of gaze and the deviation is termed ***incomitant***. The drawing below shows an incomitant esotropia with the esotropia greater in left gaze, caused by limited abduction left eye. On left gaze the left eye does not move out and the right eye turns in fully causing an ET greater in left gaze.

**Incomitant - Esotropia**



## Question:

Drawing (right) shows a patient fixing right eye and trying to looking up. The right eye has restricted elevation because the inferior rectus muscle is entrapped in a floor fracture.

Which is correct?

1. Left hypertropia is comitant
2. Left hypertropia is incomitant and is greater in downgaze
3. Left hypertropia is incomitant and is greater in upgaze
4. Drawing shows the primary deviation

## Answer:

The restriction of elevation right eye causes a left hypertropia increasing in upgaze, thus the hypertropia is incomitant and increases in upgaze so # 3 is correct. As the right eye tries to elevate against the restriction, the normal left eye overshoots up per Hering's law. The restricted right eye is fixating so the drawing is showing the secondary deviation so # 4 is wrong.

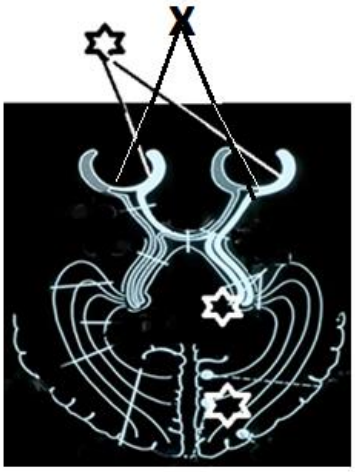
***Prism Notes:*** When using prisms to measure or neutralize a deviation do not stack them on top of each other if oriented the same direction. For example to neutralize an ET 40 PD do not place two 20 PD prisms BO on top of each other over one eye. Split them 20 PD BO each eye. You can, however, stack vertical and horizontal prisms. For example RHT 10 ET 20 can be neutralized by 10 PD BD right eye and 20 PD BO right eye.

Prisms can be ground into glasses to correct small angle strabismus. Usually a maximum of 10 PD for each eye can be used to correct a deviation up to 20 PD. Another type of prism is a Fresnel or "press-on" prism. This is a plastic sheet that has a series of small prisms in parallel which can be applied to glasses. Press-on prisms can correct up to 40 PD each eye; however, larger amounts of prism can blur vision and the patients often complain. Press-on prisms can be valuable as a temporary measure while waiting for recovery of a paralytic strabismus.

When prescribing prism glasses usually split the prisms between the two eyes, and document the orientation by the base. So to write a prescription for diplopia caused by an ET 10 PD write: RE 5 BO, and LE 5 BO. For paralytic or restrictive strabismus neutralize the primary deviation by placing the prism over the eye with limited ductions. For a right $6^{th}$ nerve paresis with an ET 20 PD left eye fixing place a 20 PD BO press-on prism over the right eye.

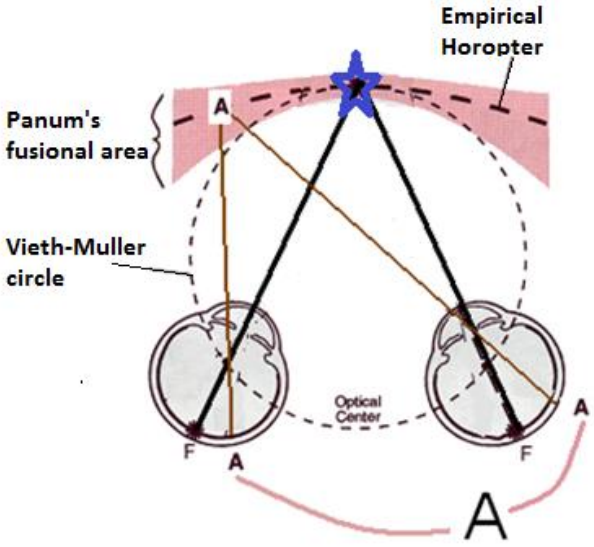
# Chapter 4 Binocular Vision and Sensory Adaptations to Strabismus

## Binocular Vision and Corresponding Retinal Points




Normally, retinal images from both eyes fuse together to give us binocular vision with stereo acuity. The merging of images occurs as axons from nasal retina of one eye cross the chiasm to join axons from temporal retina of the fellow eye. Corresponding retinal points are specific areas of nasal and temporal retina that are linked in the cortex. This is termed "Normal Retinal Correspondence". Diagram right shows the eyes fixing on an **X** straight ahead, with a six pointed star to the left. Note that the star projects to nasal retina left eye and temporal retina right eye. These are corresponding retinal points as axons from these points join by way of the chiasm, and lateral geniculate nucleus to merge in the occipital cortex where visual information is processed into binocular vision.

Diagram (lower right) shows a patient fixating on a star. The dark dashed curved line that goes through the star is the empirical horopter. Objects in this line will project to corresponding retinal points and be fused as one. Note point "**A**" peripheral to the star is on the empirical horopter so it projects to corresponding retinal points and is fused as a single image. The pink area is Panum's fusional area. This is an area where objects off the empirical horopter will still be fused. Vieth-Muller's circle is a mathematical calculation using the optical centers of the eyes to determine where corresponding retinal points should be located. Sensory testing has shown Vieth-Muller's circle to be wrong. The empirical horopter demonstrates the non-linear aspect of vision as central retinal receptive fields are smaller (high resolution) while peripheral receptive fields are larger (low resolution).



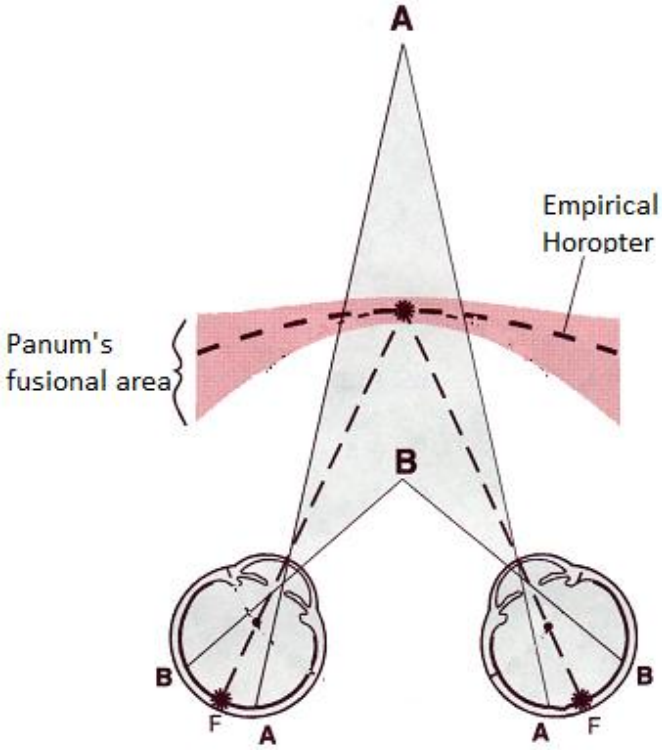
**Question:** Why do we have a chiasm?

1. It gives us binocular vision and stereo acuity.
2. It gives us consensual pupillary response.
3. So we can be quizzed on Wilbrand's knee
4. All of the above

**Answer:** The chiasm allows corresponding retinal points from nasal retina of one eye to cross and join temporal retina of the fellow eye, thus giving us binocular vision and stereo acuity. Answer # 2 "Consensual pupil" is also correct and come to think about it so is answer # 3. Neuro-ophthalmologists love to bug us about Wilbrand's knee. The answer is # 4, all of the above.

## Physiologic Diplopia

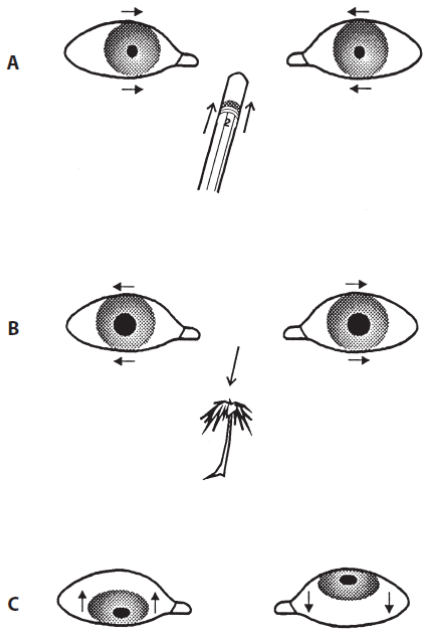
When an object is outside of Panum's fusional area the object will stimulate non-corresponding retinal points and cause double vision termed physiologic diplopia. Diagram (right) shows the patient fixating on a black dot in the center of the empirical horopter. Object A is distal to the empirical horopter so its image is projected to bi-nasal retina that are non-corresponding points. Object B is proximal to the empirical horopter and its image stimulates bi-temporal retina that are also non-corresponding points. Stimulation of the non-corresponding points will cause diplopia- physiologic diplopia.




You can experience physiologic diplopia by looking at a distant object across the room. Now hold a pencil a few inches in front of your nose. While you focus on the distant object you will see two pencils at near. Close your right eye and the left image disappears indicating crossed diplopia.

## Normal Binocular Fusion

There are two types of binocular fusion: motor and sensory.



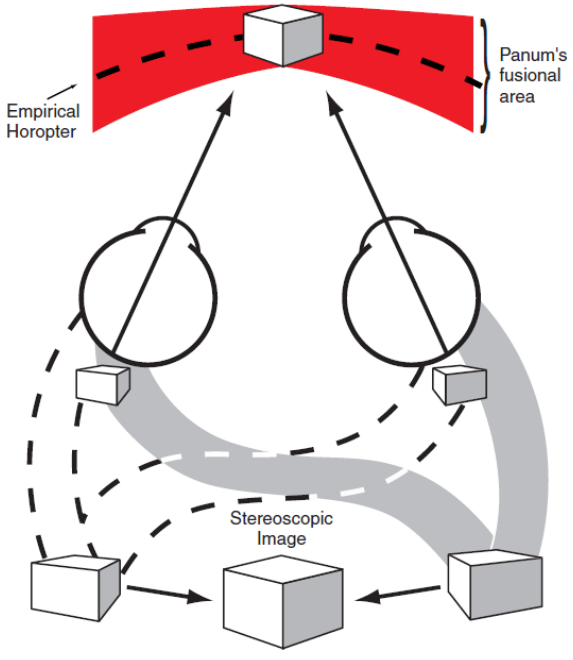

**Motor Fusion:** As described in chapter 2 we use vergence movements to adjust eye position and maintain proper alignment. This mechanism is called motor fusion. The stimulus for vergence movement is when the image moves off of corresponding retinal points. We use convergence for an approaching target (drawing A), divergence for a target moving away (drawing B), and vertical vergence to control vertical drift (drawing C). Motor fusion also can control a small strabismus keeping the eyes aligned. A strabismus controlled by motor fusion is called a **phoria**.

***Normal Vergence Amplitudes***

Convergence = 25 to 30 PD

Divergence = 6 to 8 PD

Vertical = 1 to 2 PD




**Sensory Fusion:** Three dimensional objects will not lie directly on the empirical horopter, as part of the object will be in front and behind the line. Thus three dimensional objects will stimulate noncorresponding retinal points. As long as these noncorresponding retinal points are within Panum's fusional area they will be fused and stimulate three dimensional stereoscopic vision. This fusion that gives us stereoscopic vision is sensory fusion. Drawing right shows each eye has a different angle of view and a different retinal image of the visual target (the cube). Inter-pupillary distance determines stereo acuity as the farther apart the eyes the more disparate the images and the greater the stereoscopic perception. Stereo acuity requires good eye alignment and strabismus larger than 10 PD will eliminate stereo acuity. **Normal stereo acuity** = 40 seconds arc

**Question:** As a visual target moves to distance there is a reduction in stereo acuity. True or False?

**Answer:** As an object moves away the difference in visual angle diminishes and the images to each eye become more similar. The reduction in image disparity results in a reduction in stereo acuity as an object moves away so the answer is true. Visual targets farther than 20 feet (6 meters) stimulate very little stereopsis. You can experience this phenomenon. Place your hand a few inches in front of you. Close one eye then the other and note that each eye has a different view of your hand. Now look at a distant object across the room and note each eye has a very similar view.

## Diplopia (Binocular)

Acquired strabismus in older children (> 4 to 5 yrs) and adults causes binocular diplopia or simply termed diplopia. Young children have the ability to cortically suppress the second image and typically do not experience diplopia. Diplopia occurs as one eye fixes on the target with the fovea (central image), while image in the deviated eye falls on peripheral retina and is perceived as a second image in the peripheral visual field. Diagram upper right shows esotropia and the red filter test, with the red filter over the deviated eye. The left eye is fixing on the target and the right eye is deviated so the image falls on nasal retina. Since nasal retina projects to the temporal visual field, the patient sees ***uncrossed diplopia*** with the diplopic image (red) seen to be on the same side as the red filter. It is interesting that the foveal image from the deviated eye is cortically suppressed and the image from peripheral retina is perceived as the diplopic image.

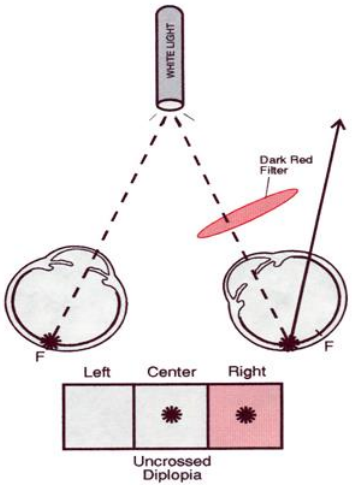
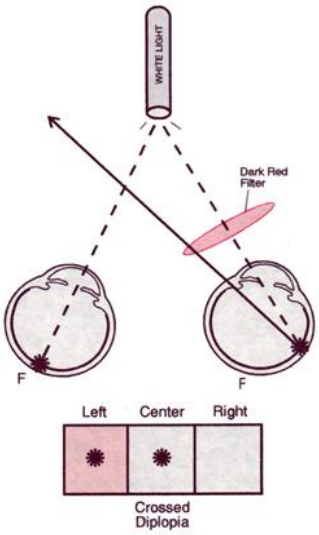



Diagram lower right shows an exotropia with the image of the target falling on temporal retina of the deviated eye which projects to the opposite visual field and causes ***crossed diplopia***.




## *Remember:*

***S in eSo is for same side - uncrossed diplopia***

***X in eXo is for a cross - crossed diplopia***

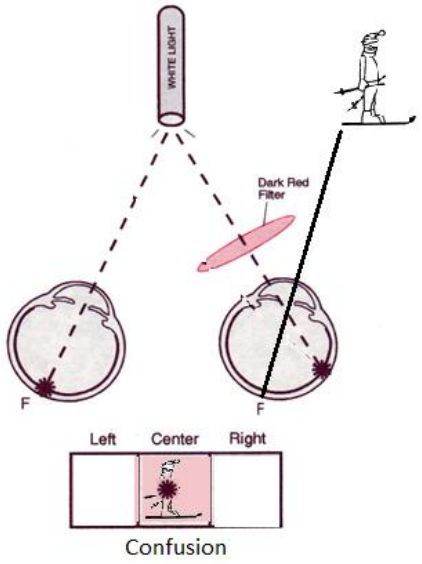
***Vertical diplopia*** causes "opposite" diplopia as the hypertropic eye will see the image lower and the hypotropic eye sees the image higher. ***Torsional diplopia*** is also "opposite" with extorsion perceived as a twisted image with the 12:00 o'clock position rotated nasally and intorsion perceived as the 12:00 o'clock position rotated temporally.

## Monocular Diplopia

Monocular diplopia is not related to strabismus, but is double vision because the image is optically split within one eye. Causes of monocular diplopia include dry eye, corneal opacity, large astigmatism, cataract, scroll of posterior capsule, or a displaced IOL. Note retinal folds and macular pucker will cause metamorphopsia, but not monocular diplopia. The image must be optically split in front of the retina to cause monocular diplopia. With monocular diplopia the second image is very blurred and often seen as a ghost image. A pin hole eliminates monocular diplopia. In general unless specified the term diplopia refers to binocular diplopia.

## Confusion

Patients with binocular diplopia cortically suppress the image from the fovea of the deviated and see two identical images: one from the fovea of the fixing eye, and the same image from the periphery of the deviated eye. Rarely, however, an adult patient with acquired strabismus will complain of seeing two different images superimposed on each other, rather than diplopia. This is called confusion, and is caused by simultaneous perception from both foveas. Figure right shows the red filter test on a patient with exotropia and sensory confusion. The patient simultaneously perceives images from both foveas and therefore sees the light superimposed on the skier. Confusion is sometimes seen in patients with end stage glaucoma and acquired strabismus due to a large implant bleb. These end stage glaucoma patients have tunnel vision so they are forced to use both foveas.



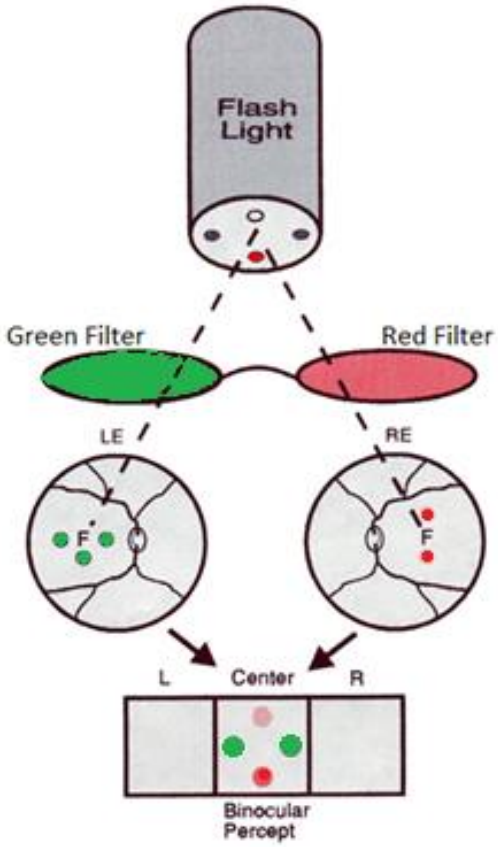

**Question:** A patient complains of vertical diplopia so you place a red filter over the right eye. The patient sees the red image to be down, and tilted with the 12:00 o'clock position nasal. Which muscle is weak?

1. Right Inferior oblique palsy
2. Left inferior oblique palsy
3. Right superior oblique palsy
4. Left superior oblique palsy

**Answer:** Let's start with torsion. The red image from the right eye is tilted with the 12:00 o'clock position subjectively nasal thus indicating anatomical extorsion. Which oblique palsy causes extorsion? The superior oblique muscle intorts so a SO palsy causes extorsion therefore possible answers are # 3 and # 4. Since the right eye is extorted the answer is probably #3 right SO palsy. We can verify this. The superior oblique muscle is a depressor so a SO palsy results in an ipsilateral hypertropia. Since the red image from the right eye is seen down that means the right eye must be up – a right hypertropia. The answer is # 3 a right superior oblique palsy.

## Sensory Tests for Binocular Vision

### Worth Four Dot Test



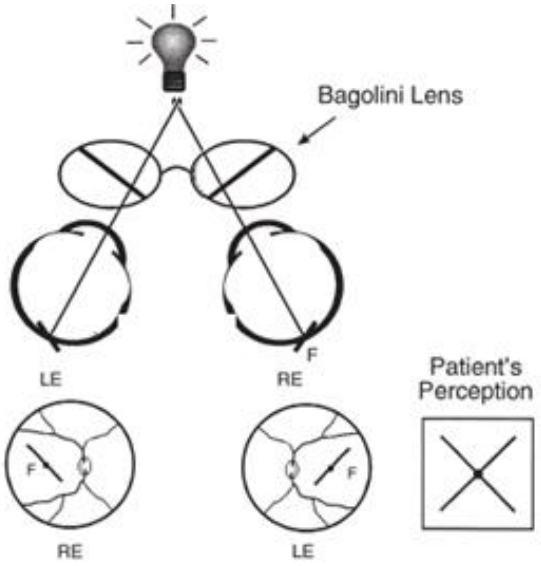

Worth four dot tests for binocular fusion. Diagram right shows the Worth four dot test. The patient wears red - green glasses then views a flashlight with four lights: one white light, one red light below, and two green lights to either side. The red filter eye sees two dots: the red dot and white dot appears red through the red filter. The green filter eye sees three dots, two green lights and the white dot filters green. The single white light is the only light seen by both eyes so it is the only binocular target. The white light is fused resulting in a normal response of seeing four dots: two green and two red. The white light actually looks kind of pinkish because it is a fusion of red and green. Patients with diplopia see 5 dots, two red and three green.

Worth four dot flash light is customarily performed at near (14 inches) which subtends a 6 degree angle, or at distance Worth four dot light box (20 feet) which subtends 1.25 degrees. Note as a visual target moves away it subtends a smaller visual angle (it appears smaller). By comparing responses from distance vs. near, Worth four dot can estimate the size of a suppression scotoma (see below).

Near stimulates peripheral field (6 degrees)

Distance stimulates central field (1.25 degrees)

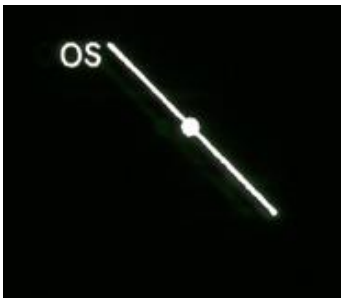
### Bagolini Striated Lenses




Bagolini lenses are clear with a scratch that produces a linear streak of light when the patient views a light source. The test is performed by placing a Bagolini lens over each eye, and having the patient fixate on a light target. The striations in the lenses are oriented at 45 degrees off the vertical meridian so the images are 90 degrees to each other. Diagram right shows Bagolini lenses used in a normal patient with binocular fusion. Note each fovea is aligned on the light so a streak of light goes through each fovea at 45 degrees, thus producing the perception of a cross. Bagolini lenses are minimally dissociating because the lenses are clear and provide a relatively unobstructed binocular view.

## Cortical Suppression

Cortical suppression is a defense mechanism to eliminate the confusion or diplopia caused by strabismus or unequal visual input from a unilateral blurred image. Children under 6 years have significant neuro-plasticity and the immature brain actually suppresses neuronal activity in the visual cortex that corresponds to the deviated eye or the blurred image. Suppression is why children with strabismus do not see double. Older children and adults lose the ability to suppress so acquired strabismus results in diplopia. Some adults, however, can learn to suppress double vision, but it can take several months to years. Cortical suppression can involve a large area of the visual field of the deviated eye (i.e. large suppression scotoma) or a small central area (central suppression scotoma) if there is a small deviation.




**Bagolini Lens:** Diagram right shows the response to Bagolini lens testing in a patient with a large angle esotropia and large area of suppression right eye. Note that the patient only sees the light streak from the fixing left eye and totally suppresses the image from the deviated right eye.

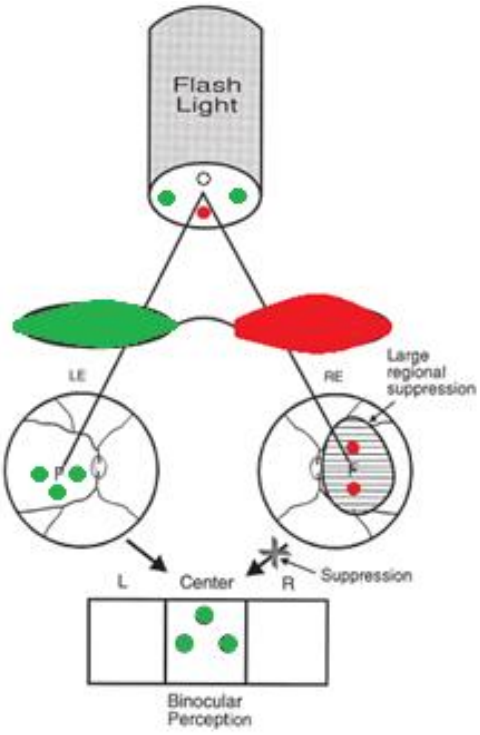
## Strabismus Induced Suppression

There are 3 patterns of suppression associated with strabismus.

- ***Large regional suppression*** - no fusion and no stereo acuity
- ***Central suppression - peripheral fusion (Monofixation Syndrome of Parks) –*** some fusion with low grade stereo acuity
- ***Anomalous retinal correspondence (ARC) –*** superimposition of images but no fusion and no stereo acuity

## Large regional suppression (no fusion)

Suppression areas larger than 5 degrees (10 PD) in general preclude useful binocular fusion and stereo acuity. Patients with large regional suppression are basically monocular when they are tropic and cortically suppress visual information from the deviated eye. They do not have diplopia. Patients with childhood intermittent strabismus can have both excellent binocular fusion with high grade stereo acuity and large regional suppression. When they are aligned (orthotropia) they have excellent stereo acuity, but when they are tropic they cortically suppress the image from the deviated eye and have no stereopsis. This ability to suppress and fuse is seen in intermittent exotropia. These patients have excellent stereo acuity of 40 sec arc when orthotropic and fusing, but have large regional suppression and no stereo acuity when the exotropia is manifest (see Chapter 8, P-96). Patients with large angle constant esotropia will not fuse. Some with equal vision will alternate fixation and suppression, while others have strong fixation preference for one eye, and always suppress the same eye. In young children, strong fixation preference with constant suppression of one eye is associated with amblyopia of the suppressed eye.




Large regional suppression results in suppression of both near and distance Worth four dot as the suppression scotoma is larger than the angle subtended by Worth four dot. Near Worth four dot is shown figure right in a patient with esotropia of 20 PD and corresponding suppression scotoma of 10 degrees. Note the patient only sees dots in the fixing eye and suppresses the two dots in the deviated eye.

**Remember: 1 degree = 2 prism diopters**

## Monofixation Syndrome of Parks

Patients with a small angle esotropia less than 10 PD can develop binocular fusion of the peripheral retinas, yet suppress the central field of the non-preferred eye (see diagram of Worth four dot below). This was first described by the late Dr. Marshall Parks. These patients have motor fusion and some low grade stereo acuity between 60 and 3000 seconds arc. Monofixation is sometimes called the tropia phoria syndrome as these patients can have a small tropia (<10 PD) and a larger phoria. This is diagnosed by cover-uncover test that will show a small esotropia, then alternate cover testing discloses a larger esophoria. Patients with monofixation syndrome often have amblyopia but it is usually mild. Hyperopic anisometropic amblyopia can cause the monofixation syndrome with a central suppression scotoma in the hyperopic eye.

**Worth four dot** is an excellent way to diagnose monofixation syndrome. Diagram right shows monofixation syndrome with peripheral fusion and a 4 degree central suppression scotoma left eye. The distance Worth four dot is being used that subtends 1.25 degrees and stimulates the central field. Note that the two red dots fall within the central suppression scotoma so they are not seen. Only the three green dots from the fixing eye are perceived.

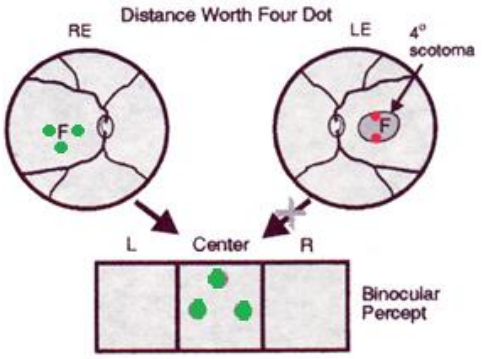
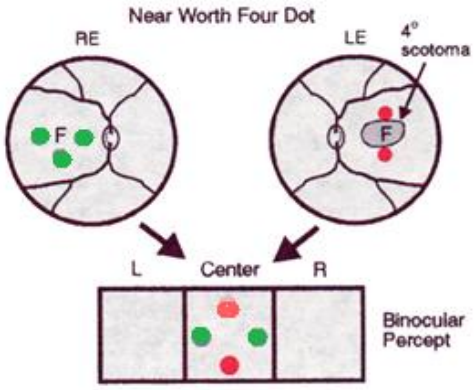
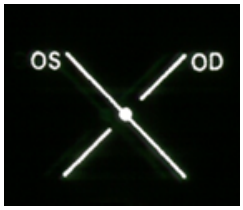



Diagram right shows the near Worth four dot subtending 6 degrees and stimulating the peripheral retina. Note the two red dots are falling outside the suppression scotoma so the patient sees four dots indicating peripheral fusion.




*This pattern of suppressing the distance Worth four dot and fusing the near Worth four dot is virtually pathognomonic for monofixation syndrome and is often a board question.*

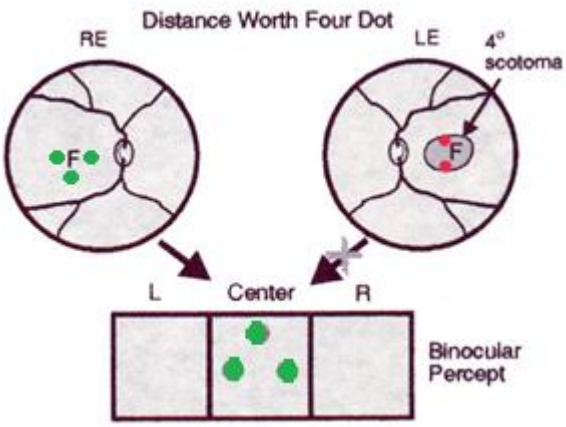
**Bagolini lenses:** Diagram right shows the perception of a patient with small esotropia with monofixation syndrome (central suppression and peripheral fusion) examined by Bagolini lenses. Note the Bagolini line striations are seen by both eyes indicating peripheral fusion, and the missing center aspect of the OD line indicates a central suppression scotoma right eye.

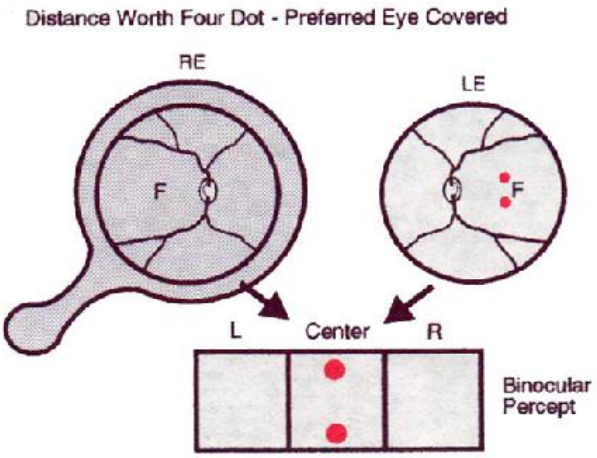


## Conditions associated with monofixation syndrome:

- Small angle esotropia (<10 PD) usually after surgery for congenital ET
- Anisometropic amblyopia
- Primary monofixation syndrome unknown cause (rare)
- Foveal hole or scar has also been historically included because these patients have a central scotoma and peripheral fusion. However, in the case of foveal pathology there is an absolute scotoma, it is not a suppression scotoma that does not disappear when the dominant eye is occluded (not facultative, see below) so foveal pathology probably should not be classified as monofixation.

**Facultative Scotoma:** A suppression scotoma is facultative. That is, it only exists during binocular viewing. Covering the fixing eye eliminates the suppression scotoma. Suppression is present to eliminate the disparity produced by unequal visual inputs from strabismus or a unilateral blurred image. You occlude the fixing eye, you eliminate the competition, and you eliminate the suppression. The diagram below left depicts monofixation syndrome with a central suppression scotoma left eye, so the patient sees three green dots and suppresses the two red dots on Worth four dot testing. Diagram below right shows that occluding the fixing eye (RE) eliminates the central suppression scotoma of the non-dominant (LE) eye thus causing the patient to see two red lights.



### Question:

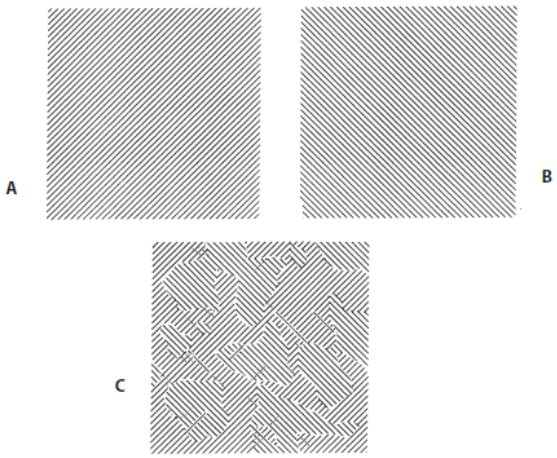
A five year old presents after having strabismus surgery at 8 months of age for congenital esotropia. Cover uncover test shows an esotropia 6 PD and alternate cover an esophoria of 12 PD. Worth four dot would show:

1. Three dots at distance and near
2. Four dots at distance and near
3. Three dots at distance and four dots at near
4. Four dots at distance and two at near

**Answer:** Cover-uncover test identifies a small esotropia of 6 PD. Alternate cover dissociates fusion and brings out the latent phoria so the deviation increases to 12 PD. The presence of a small tropia and a phoria indicates monofixation syndrome (peripheral fusion central suppression). These patients suppress the distance Worth four dot that stimulates 1.25 degrees that is within the central scotoman caused by the tropia. The near Worth four dot is fused as it subtends 6 degrees which is outside the central scotoma. The answer is # 3, three dots for distance (central suppression) and four dots for near (peripheral fusion).

## Retinal Rivalry

Different images falling on corresponding areas of the macula result in patchy local suppression of the image so there is no simultaneous perception of different images. Diagonal lines (figures A & B) presented to each eye's macula oriented 90 degrees to each other produces different images to corresponding retinal points. This results in patchy cortical suppression of local areas of the macular visual field and is termed retinal rivalry (figure C). Retinal rivalry requires binocular fusion.

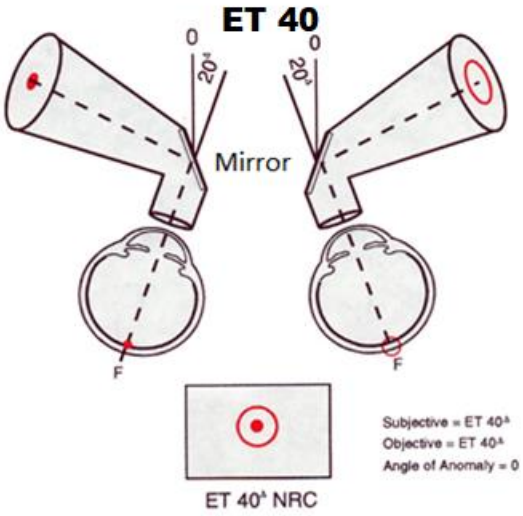



## Normal Retinal Correspondence

Normal retinal correspondence (NRC) is the normal state where the visual cortex recognizes the fovea of each eye as the visual center and images falling on nasal retina are cortically fused with images on corresponding temporal retinal, (see P. 44). A patient with NRC and esotropia will see single if the image in the deviated eye is moved to the fovea either by prism, strabismus surgery or a haploscopic device (see below).

## Haploscope Device

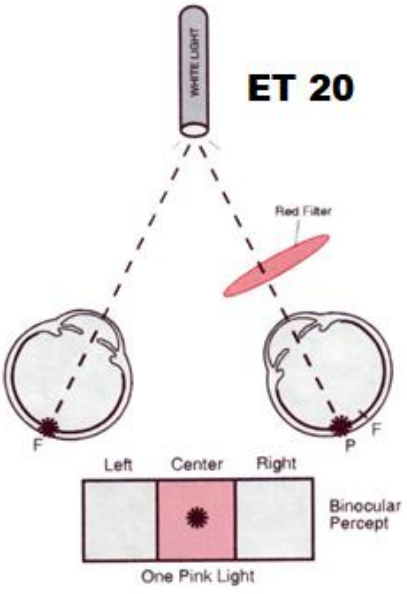
A haploscopic instrument allows presentation of different images to each eye (drawing right). This virtually splits vision in half thus the term haploscopic. The synoptophore (major amblyoscope) and troposcope are haploscopes and have mirrors that can be angled to project images to each eye. The drawing right depicts an esotropia of 40 PD objectively measured by alternate cover test. The patient subjectively angles the mirrors so the dot is placed in the center of the ring. If there is NRC angling the mirrors will place the dot on one fovea and the circle on the other fovea wto make a bullseye.



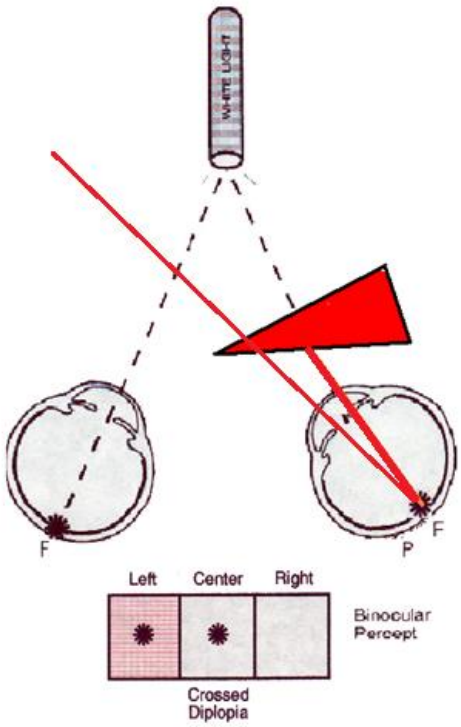

In the drawing the mirrors are angled of 20 PD each mirror total of 40 PD. When the subjective angle is the same as the objective angle this indicates **normal retinal correspondence** as the true foveas are used as the visual center. Anomalous correspondence (ARC) the subjective angle will be less, even zero because they use the pseudo fovea, not the true fovea of the deviated eye (see below).

## Anomalous Retinal Correspondence

Some children with strabismus develop a curious sensory adaptation to eliminate diplopia and confusion termed anomalous retinal correspondence (ARC). With ARC the visual cortex adapts to strabismus by reorientation so the visual center of the deviated eye moves from the true fovea to a peripheral retina point where the diplopic image falls called the pseudo-fovea. The drawing right shows a right esotropia with the retinal image of the deviated eye falling on nasal retina. This patient has ARC so the brain has reoriented making this nasal retinal point where the diplopic image lands the new visual center, i.e. the pseudo-fovea (P). The pseudo-fovea corresponds to the true fovea of the fixing eye so the patient does not see double. ARC patients do not actually have binocular fusion. There is neither motor fusion nor stereo acuity. ARC patients just superimpose the image from the pseudo-fovea and the true fovea to eliminate diplopia.

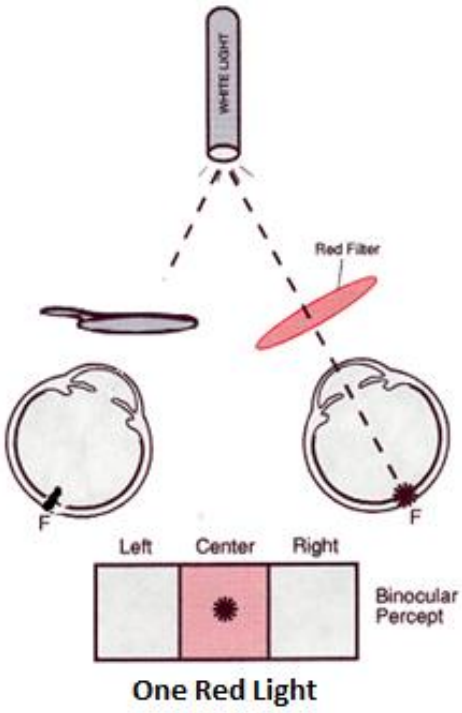


**ARC and Paradoxical Diplopia:** If the image is moved off the pseudo-fovea with a prism, strabismus surgery or a haploscopic device you will induce diplopia. Diagram right shows the patient with esotropia and ARC. A base out prism is placed over the deviated eye to move the retinal image off the pseudo-fovea onto the true fovea. This neutralizes the esotropia, but creates crossed diplopia because the image is now temporal to the pseudo-fovea. Retina located temporal to the pseudo fovea is considered as falling on temporal retina and will cause crossed diplopia. This diplopia is called **paradoxical diplopia** because the patient sees double when the deviation is corrected.



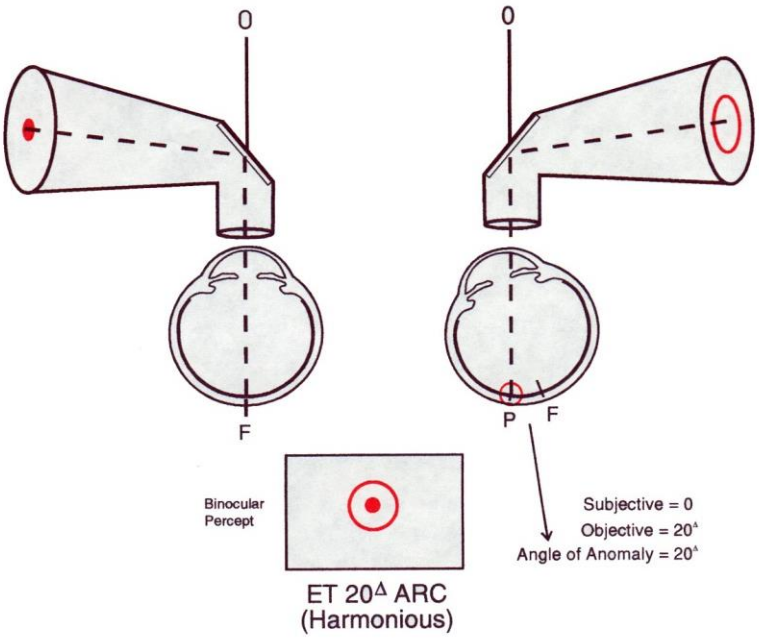

**Pseudo Fovea disappears during Monocular Viewing:** ARC and the pseudo fovea goes away when you cover the fixing eye as the patient will now fixate with the true fovea. Covering one eye eliminates the strabismus and need for a pseudo fovea. ARC and the pseudo-fovea are only needed to eliminate diplopia during binocular viewing and disappear when you cover one eye.

Figure right shows the patient with ARC but the left dominant eye is occluded. Occluding the dominant eye caused the pseudo-fovea to disappear and the true fovea now fixates.



## Haploscopic Device and ARC

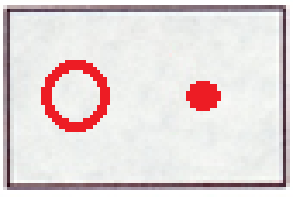
A haploscopic device can be used to study ARC. The diagram below shows esotropia with ARC. The patient directed to angle the mirrors to put the red dot in the center of the red circle. Because the patient uses the pseudo fovea under binocular viewing and the pseudo fovea is in line with the target (red circle) the mirrors are not moved. This is the subjective angle and in this case is zero.



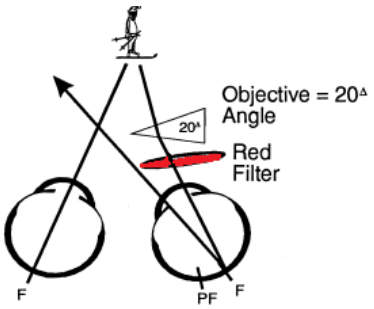

The objective angle is measured by alternate cover testing and in this case is 20 PD. Alternate cover testing keeps one eye covered so the pseudo fovea disappears and the true foveas are used. Objective angle is the true fovea to true fovea deviation.

The difference between the subjective and objective angle is the **Angle of Anomaly** and is the measurement of the eccentricity of the pseudo fovea. If the subjective angle is zero this means the pseudo fovea is in line with the deviated eye and objective angle will be the same as the angle of anomaly. In the example above the angle of anomaly is 20 PD the same as the objective angle and this is termed **harmonious ARC**. Harmonious ARC means the pseudo fovea location matches the deviation. If the subjective angle is not zero then the pseudo fovea location is not match the deviation and this is termed **unharmonious ARC.**

***Note:*** Covering one eye forces fixation to the true fovea, so alternate cover testing measures the objective angle – the true fovea to true fovea deviation. If the mirrors above are angled based on alternate cover testing to neutralize the deviation they would angle 20 degrees. This angulation moves the circle off the pseudo-fovea to the true fovea and would cause crossed diplopia with the circle on the left of the dot as shown in drawing to the right (paradaoxical diplopia).



**Postoperative Paradoxical Diplopia:** Surgical correction of strabismus in adults with ARC can result in paradoxical diplopia. It is therefore important to test for ARC prior to adult strabismus surgery.

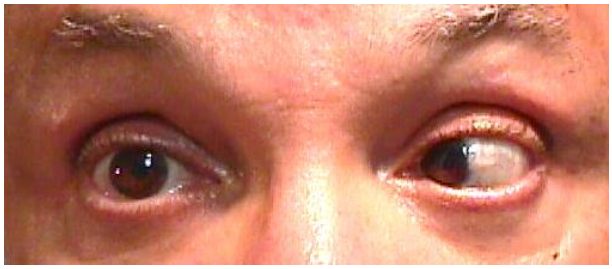




Clinically the most useful test for ARC and possible postoperative diplopia is prism neutralization with or without a red filter (see drawing right). Measure the deviation with alternate prism cover testing and correct the deviation with the appropriate prism. If prism neutralization results in diplopia there is ARC and it is likely the patient will have postoperative diplopia. If the patient does not see double with prism neutralization then it is highly unlikely they have ARC and tells us postoperative diplopia should not be a problem.

Even if paradoxical diplopia is found on prism neutralization many patients are not bothered by this as they know which is the real image, and they ignore the diplopic image. If not overly bothered it is usually ok to proceed with strabismus surgery. Uncommonly patients will be extremely uncomfortable with diplopia after prism neutralization. If this is the case, it is probably better not to operate, and *just refer this patient to your competition down the street*. In any case, it is always prudent to advise adult patients that they might see double after strabismus surgery (Oh - and document this in the chart).

**Question:** This 40 year old has a long standing esotropia measured at 50 PD by prism alternate cover testing. What test result indicates if the patient has ARC and will likely see double if the esotropia is surgically corrected?

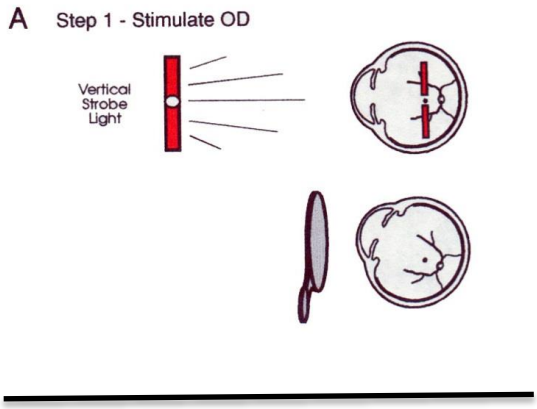


1. Prism neutralization 50 BO causes diplopia
2. Synoptophore angled for ET 50 PD causes diplopia
3. Red filter test shows no diplopia
4. Answers # 1 and # 2

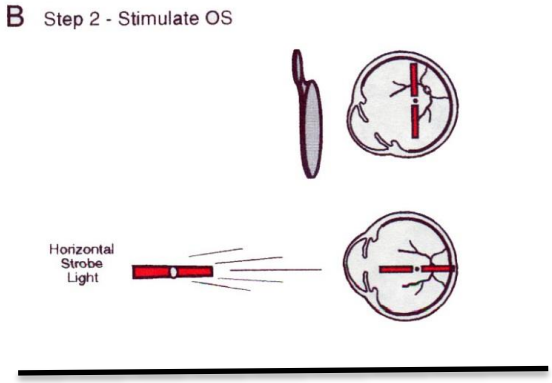
**Answer:** Both prism neutralization with 50 BO, and angling the synoptophore (a haploscopic device) for ET 50 PD places the images on both true foveas. If placing the images on the true foveas causes diplopia this “paradoxical” and indicates the presence of a psedo-fovea and ARC . These patients will likely have post operative diplopia and the answer is # 4 as both # 1 and # 2 are correct. Answer is # 3 is wrong as no diplopia with red filter could be ARC or large suppression, not specific for ARC.

## Afterimage Test

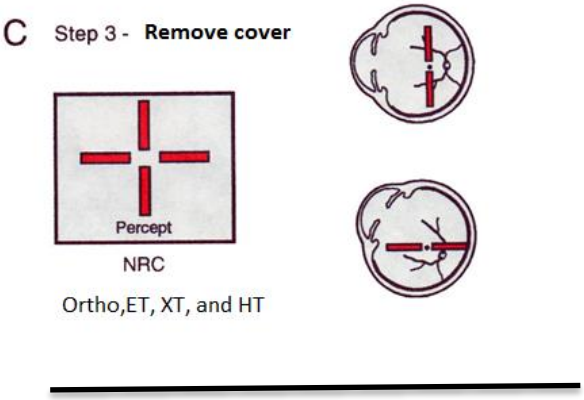
Afterimage test is based on tagging the true fovea of each eye with a bright strobe light that leaves an afterimage. This is done by occluding one eye and having the patient look at a vertically oriented tubular strobe light masked in the middle (see figure A). A flash of the strobe creates a vertically oriented afterimage centered on the fovea.



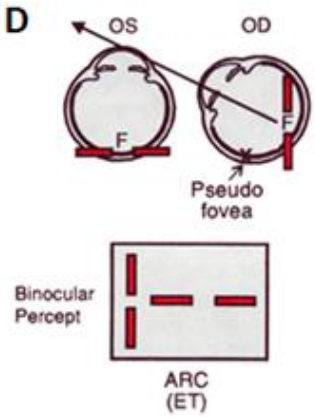

In drawing B the occluder is moved to the fellow eye. The strobe is now horizontally oriented to produce a horizontal after image centered on the other eye (Figure B).




**Normal Retinal Correspondence (NRC):** Figure C shows the occluder removed and the subjective response of a patient with NRC. Since the strobe has placed an afterimage on each fovea, note that the patients with NRC see a cross regardless of the presence or absence of strabismus or if they are orthotropic. NRC patients always orient their visual center to their true fovea.

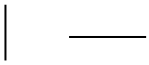



**Anomalous Retinal Correspondence (ARC):** Figure D shows the afterimage test on a patient with esotropia and ARC. Note that both true foveas are tagged because the strobe was applied under monocular conditions. When both eyes are opened the patient regains the ARC and pseudo-fovea. Since the pseudo-fovea is considered the visual center, the tagged true fovea is perceived as eccentric. Strabismic patients with ARC will never see a cross.



## Question:

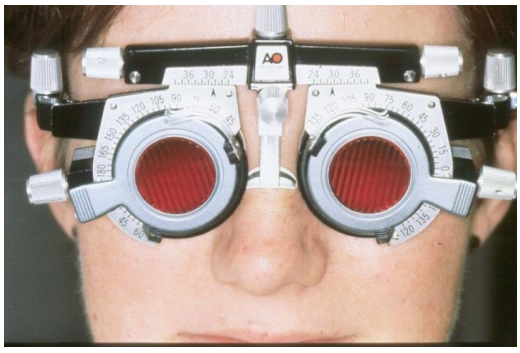
An adult patient has an esotropia of 20 PD and prism neutralization with 20 PD BO gives the patient crossed diplopia. Afterimage test is given and the most likely response is:

1. A cross afterimage 
2. Lines of the afterimage side by side (crossed) 
3. Vertical offset of the after image
4. Suppression of one of the afterimages

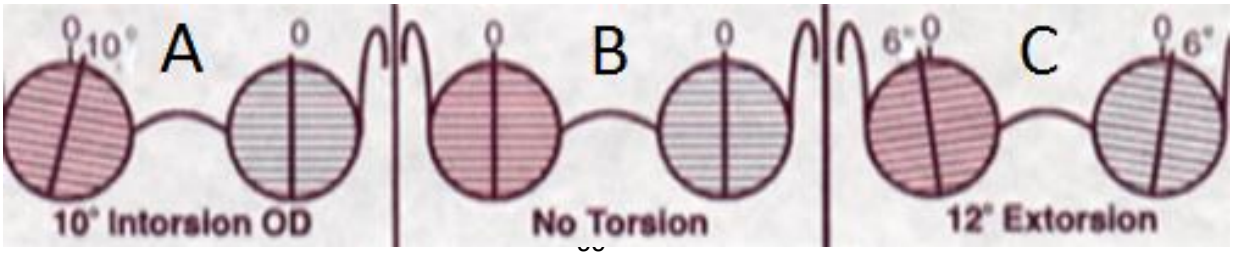
**Answer:** Since prism neutralization produced crossed diplopia (paradoxical diplopia) this indicates the patient has ARC. Afterimage is stimulated under monocular conditions so each fovea is tagged no matter what the eye alignment or sensory adaptation so patients with NRC always see a cross. Patients with ARC see the afterimages offset because the foveas are tagged and after tagging with both eyes are opened so the pseudo fovea becomes active and is the visual center. The tagged true fovea of the deviated eye temporal to the pseudo fovea thus the perception of side by side lines - so #1 is wrong and #2 is correct. Answers #3 is wrong as the deviation is horizontal and # 4 is wrong as the patient has ARC, not large regional suppression.

## Maddox Rod

Maddox rod (photo right) is a series of high power plus lenses in parallel orientation that produces a line of light when the patient views a light. The line of light is 90 degrees to the parallel lenses. The Maddox rod is the most dissociating sensory test as the patient only sees a line of light without any other fusion clues. It will disclose even small phorias. Double Maddox rods can be used to diagnose torsion. The patient rotates the lens so the line of light is vertically straight up and down or straight horizontally.



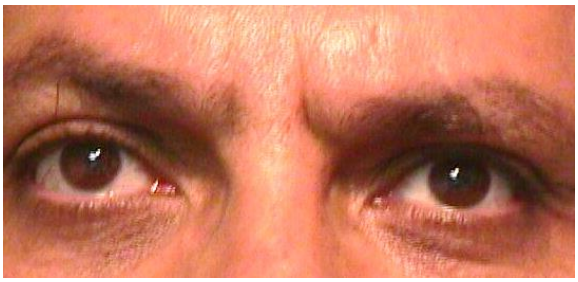
Below figure A shows right intorsion (12:00 o'clock tilted nasally), figure B shows no torsion with the line vertically parallel, and figure C shows each eye extorted 6 degrees for a total of 12 degrees extorsion.



## Dissociating Binocular Fusion

Sensory tests can break down binocular fusion by causing asymmetric visual input, and these tests are called dissociating tests. An occluder over one eye is most dissociating and is why alternate cover test dissociates fusion and discloses a phoria. In the dark the Worth four dot test is a very dissociating test, because the sole binocular fusion clue is the one white light. With the room lights on the patient can see through the red green glasses and see the room content including the examiner which provides peripheral binocular fusion clues. Patients with a phoria will often be dissociated with Worth four dot done in the dark. Except for an occluder, the Maddox rod is the most dissociating test as the image is distorted and all the patient sees is a streak of light. Bagolini testing is least dissociating because there is a clear view for both eyes through the clear lens.

**Question:** An adult patient is complaining of intermittent horizontal diplopia. There is no shift on cover uncover testing and stereo acuity measures 40 seconds arc. What test or tests would best help make the diagnosis?



1. Bagolini lenses
2. Alternate cover test
3. Maddox rod
4. Answers #2 and #3

**Answer:** The patient has good alignment, stereo acuity and no tropia by cover uncover testing, but intermittent diplopia so there is a possibility of a latent deviation, i.e. a phoria. To show a phoria we must break down fusion and this is done with dissociating tests. Bagolini lenses are clear and are least dissociating so answer # 1 is wrong. Alternate cover testing and Maddox rod testing are both very dissociating so the answer is # 4.

## Horror Fusionis

Horror fusionis is the rare strabismic condition where there is an inability to fuse and suppress so the patient has intractable diplopia. These patients lack binocular fusion so they cannot fuse the images, yet they cannot suppress so they have diplopia. Correction of the strabismus with prisms or strabismus surgery results in superimposition of images but the patient still has double vision.

Clinically horror fusionis can occur in adults with strabismus who lose vision in the dominate eye (e.g., traumatic cataract). Rehabilitation of vision is delayed (usually > a year) so the non-dominate eye is no longer suppressed and shares dominance. When the vision is restored in the dominate eye the patient cannot fuse and cannot suppress so they are left with intractable diplopia – horror fusionis.

**Anti-suppression therapy** can also cause horror fusionis. Anti-suppression therapy has been used to treat childhood strabismus by reducing suppression and teaching the child to use the non-dominant eye. Anti-suppression therapy actually trains patients to see diplopia. It is usually done by placing a red filter of increasing density over the dominant eye until the non-dominant eye sees a light. At this point the patient will see double, the red light from the dominant eye, and a white light from the non-dominant eye. Anti-suppression does not work to correct strabismus and should not be used because of the danger of causing horror fusionis.

## Central Fusion Disruption

Patients with severe cortical damage such as from head trauma or cerebral vascular stroke can lose binocular fusion potential and will be left with intractable diplopia termed central fusion disruption. These are patients have damage to fusional areas in the brain so they lose the ability to fuse and develop a variable strabismus with intractable diplopia.

### Treatment of Horror Fusionis and Central Fusion Disruption

Monocular occlusion is the only definitive treatment for the diplopia. Prisms and strabismus surgery to align the eyes can make the diplopia worse as the images are on top of each other. In fact, moving the images farther apart with prisms or even strabismus surgery can improve the symptom of diplopia. After a trial of prisms this author actually operated to make the strabismus worse to separate the images in a patient with horror fusionis with a happy patient as the result.

**Question:** A 40 year old develops a dense cataract after trauma to his dominant eye. After a year he develops an exotropia and is now fixing with the previously non-dominant eye. Red filter test show crossed diplopia and diplopia persists with prism neutralization. You are planning cataract surgery. What should you tell the patient prior to cataract surgery?

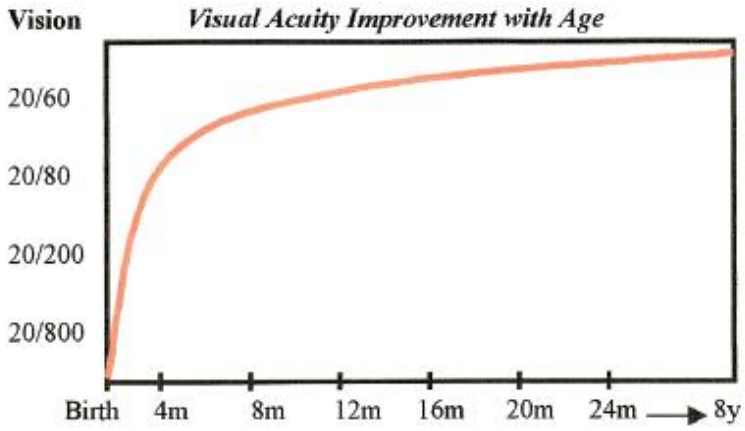
1. No problem I'm an ace cataract surgeon.
2. Thank goodness you came to me rather than to those losers down the street.
3. Can you pay cash?
4. There is a good chance you will have persistent double vision and in some cases it does not go away even after strabismus surgery or prisms and this is called "Horror Fusionis".

**Answer:** This patient is at risk for horror fusionis and diplopia so the answer # 4. Note, the other answers aren't so bad either- LOL.

# Chapter 5
# Visual Development and Amblyopia

## Normal Visual development

At birth our vision is poor. *Clear retinal images are required to stimulate development of high resolution neuro networks in the visual centers of the brain.* Normally visual acuity improves rapidly in the first few months of life called the *critical period* of visual development.




**Time Table of Visual Development (see graph above)**

- **Critical period** of visual development = first 2 months of life.
- Visual development most rapid first 2 to 3 years but plasticity continues up to 7 to 8 years of age
- Extended plasticity though adulthood but is minimal

**Disruption of Visual Acuity development**

- Unilateral or bilateral blurred retinal image
- Strabismus with constant cortical suppression of deviated eye

Amblyopia is poor vision caused by abnormal visual stimulation during the sensitive period so that neuro networks in visual areas of the brain are damaged. Abnormal simulation can be from a blurred retinal image (unilateral or bilateral), or from strabismus with constant cortical suppression of the non-dominant eye. Amblyopia is usually defined as at least 2 lines difference in best corrected visual acuity, not caused by an organic abnormality such as an optic nerve disorder or retinal disease. A child is most susceptible to developing amblyopia during early infancy but amblyopia can occur up to 5 to 6 years of age.

A severely blurred retinal image either unilateral or bilateral (e.g. dense congenital cataract) during the critical period can cause dense amblyopia and permanent visual loss if not cleared promptly.

## Pathophysiology of Amblyopia

Basic science studies have shown pathological damage associated with amblyopia involves both the lateral geniculate nucleus (LGN) and striate cortex.

Images right show a cross section of LGN. Left image is a normal LGN with six nuclear layers. Center and right images depict abnormal LGN from dense pattern distortion amblyopia with only 3 nuclear layers from the good eye and they are abnormally thick. There is neuronal atrophy of 3 layers from the amblyopic eye. (From Hubel & Wiesel, Nobel Prize 1981)

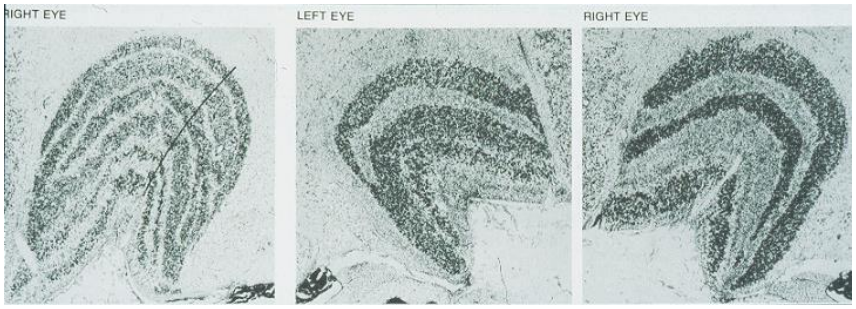
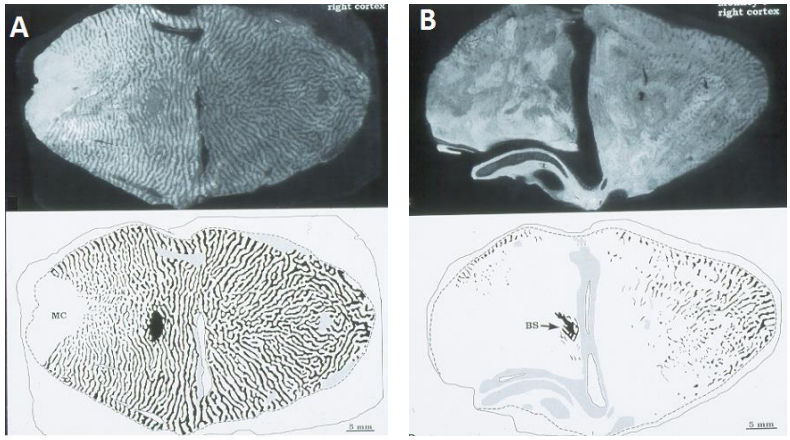



Image right (A) shows the normal zebra stripes of the striate cortex ocular dominance columns. Image (B) shows loss of ocular dominance columns from dense pattern distortion amblyopia amblyopia. (From Horton and Hocking, 1997)




**Question:** Which statement is true?

1. Amblyopia is always unilateral.
2. Amblyopia is "brain damage" in visual centers from abnormal visual stimulation.
3. Bilateral symmetrically blurred images in infancy does not cause amblyopia.
4. The critical period of visual development is the first year of life.

**Answer:** Abnormal visual stimulation during the early sensitive period causes brain damage in visual areas and is the pathophysiology of amblyopia. The answer is # 2. NOTE: Answers # 1, # 3, and # 4 are wrong because: bilateral amblyopia occurs if both eyes have a severely blurred retinal image during early visual development, and the critical period of visual development is the first 2 months of life.

## Clinical Forms of Amblyopia

There are 2 basic forms of amblyopia; **strabismic amblyopia** caused by constant cortical suppression of the deviated eye and **pattern distortion amblyopia** caused by a unilateral or bilateral **blurred retinal image** in young children.

### Strabismic Amblyopia

**Incidence**

- Congenital ET (40%)
- Accommodative ET (25%)
- Intermittent XT (2% - same as general population)

Strabismic amblyopia occurs in children with strong fixation preference for one eye and a constant deviation of the fellow eye. Patients who alternate fixation will not develop strabismic amblyopia. Note that if the esotropic patient in the photo upper right always fixates with the right eye, the deviated left eye will develop amblyopia.

### Pattern Distortion Amblyopia

**Monocular blurred retinal image** can interfere with visual development and cause amblyopia. The table below list causes of a monocular blurred image in childhood. Anisometropic amblyopia is the most common type of amblyopia with an incidence of 3 to 4 percent of the general population. It is caused by a refractive error in one eye.

| Anisometropic Amblyopia | Media Opacity |
|---|---|
| Hyperopia > 1.50 (common) | Infantile Cataract |
| Astigmatism > 2.00 | Corneal (e.g., Peters anomaly) |
| Myopia > -3.00 (unusual) | Vitreous hemorrhage |

**Bilateral blurred image** may cause bilateral amblyopia. Ametropic amblyopia is bilateral amblyopia caused by high hyperopia usually over + 5.00. Bilateral media opacity can also cause bilateral amblyopia.

| Ametropic Amblyopia | Media Opacity |
|---|---|
| Hyperopia > 5.00 | Cataract (early onset) |
| Astigmatism > 3.00 | Corneal (e.g., Peters anomaly) |

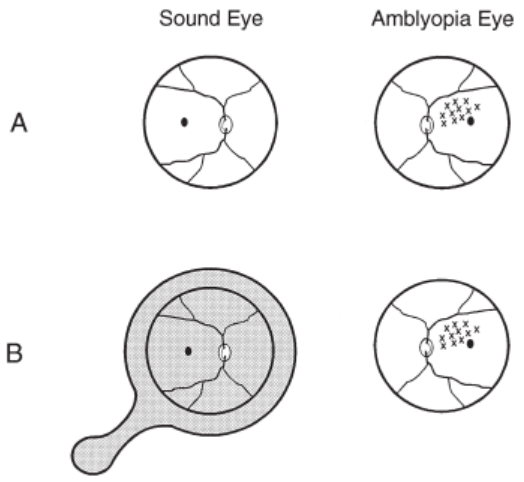
Meridional amblyopia is the term for amblyopia caused by astigmatism, either monocular or binocular.

**Sensory Nystagmus** – Bilateral severe blurred retinal images during the critical period of visual development will cause bilateral amblyopia and sensory nystagmus. This is most often associated with bilateral dense media opacity such as bilateral congenital cataracts. If bilateral visually significant congenital cataracts are not removed and aphakic contact lenses placed by 2 months of age, 90% of the patients will develop severe bilateral amblyopia and sensory nystagmus. Sensory nystagmus usually means poor vision of 20/200 OU or worse.

## Preverbal Children

For children who are too young or too uncooperative to obtain an optotype visual acuity, use monocular fixation testing to estimate vision. Cover one eye and have the patient fix and follow an accommodative target. If the child cannot fixate directly on the target or has trouble accurately following a moving target this indicates poor vision. Note the child in the photo right is looking at the target with the right eye. To assess vision move the target slowly observing how accurately the child follows the target. If the patient accurately fixes and follows the vision is good, around 20/30 to 20/80, but if fixing and following is poor, this indicates decreased vision is usually around 20/100 or worse.

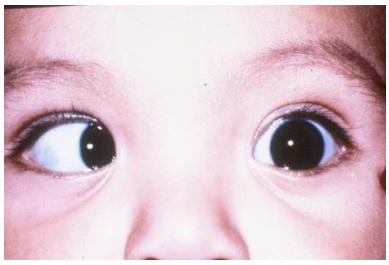
**Eccentric fixation** is very poor fixation so the patient cannot look directly at the target. Eccentric fixation indicates dense amblyopia with vision of 20/200 to count fingers and a poor prognosis. It is associated with extrafoveal viewing as foveal fixation did not develop. Drawing A shows central fixation right eye and extrafoveal viewing associated with eccentric fixation left eye. Drawing B shows that even when the sound eye is covered the patient still cannot fixate with the amblyopic fovea and continues to eccentrically view.

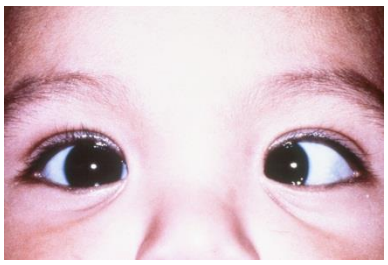



**Amblyopia - Eccentric Fixation LE**

**Note:** The pseudo-fovea associated with ARC, and the suppression scotoma associated with monofixation syndrome will disappear when the fixing eye is covered and the non-dominant eye will re-fixate with the fovea. This is not the case with eccentric fixation and dense amblyopia. With eccentric fixation, foveal fixation never developed so the fovea cannot fixate, even when the dominant eye is covered.

## Fixation Preference Testing





Fixation preference testing evaluates the ability for the child to use either eye. It adds to the information obtained by monocular fixation testing. If the patient alternates fixation this means equal visual preference and no amblyopia. Note patient right alternates fixation indicating no amblyopia.

BTW - *No! I didn't flip the slides!*

Some children will have a preferred eye but this does not necessarily mean they have amblyopia as they may occasionally alternate fixation. To diagnose strabismic amblyopia occlude the dominant eye to force fixation to the non-dominant eye, then take the cover away. If the child holds fixation with the non-dominant eye for a few seconds, through a blink, or follow a moving target before re-fixating to the dominant eye there is no significant amblyopia. If the child can't hold fixation with the non-dominant eye, and immediately re-fixates to the dominant eye this indicates significant amblyopia that should be treated.

**Question:** The child below shows preference for the left eye (left photo). You cover the dominant left eye and force fixation to the non-dominant right eye (center photo). Take the occluder away and the patient immediately re-fixates back to the left eye (photo right).

Which of the following are correct?

1. No significant amblyopia
2. Mild amblyopia left eye
3. Significant amblyopia right eye
4. Cannot tell, let's wait until the child is older and can read letters!

**Answer:** The photos above show strong fixation preference for the left eye indicating amblyopia right eye. When the dominant left eye is covered (center photo), yes the right eye will take up fixation, but as soon as the cover is removed the patient immediately re-fixates to the left eye (right photo). This indicates strong fixation preference for the left eye and significant amblyopia right eye so answer # 3 is correct.

**Note:** Early diagnosis and treatment of amblyopia is the key for obtaining good outcomes. Waiting until the child can read letters (answer 4) delays treatment and is definitely the wrong answer. That is the kind of wrong answer that can have you retaking the oral boards!

## Vertical Prism Test (Induced Tropia Test)

Fixation preference can be evaluated in patients with straight eyes by using a prism to induce strabismus. Place a 10 to 15 PD prism vertically over one eye to induce a hypertropia then evaluate fixation preference. If the patient alternates fixation or can hold fixation well with either eye there is no amblyopia. Strong fixation preference for one eye indicates amblyopia of non-preferred eye.

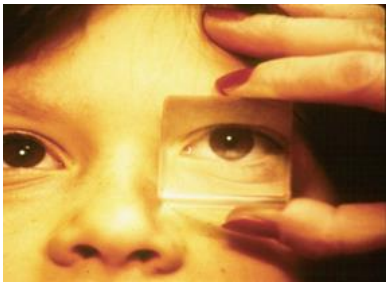
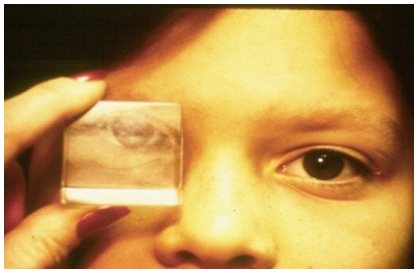


Photo upper right shows a 15 PD base down prism over left eye. The left eye is fixing through the base down prism so the eye moves up to pick up fixation. See that both eyes go up per Hering's law of yoke muscles. You can tell the left eye is fixing because the eyes move in response to the prism.



In photo lower right the base down prism is moved to the right eye. Note that the patient continues to fixate with the left eye as the left eye is looking straight ahead. This patient shows strong fixation preference for left eye and suggests amblyopia right eye.

*The vertical prism test works well for diagnosing amblyopia in patients with small angle esotropia and straight eyes, however, patients with intermittent exotropia often have strong fixation preference without amblyopia.*

Personal Note: I developed the vertical prism test with orthoptist Francis Walonker when I was a second year resident –

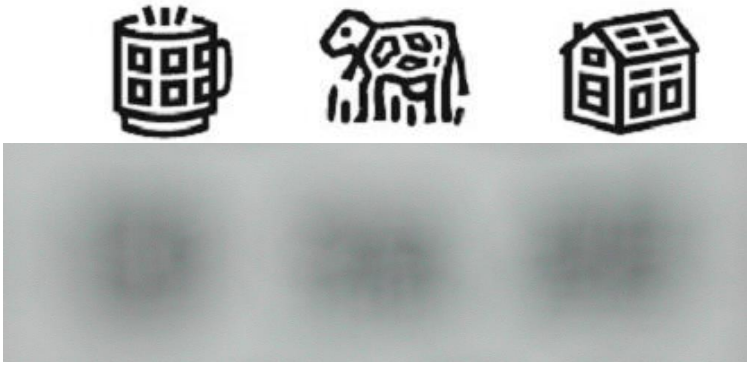
Wright KW, Walonker F, Edelman P. The 10-diopter fixation test for amblyopia. Arch Ophthalmol 1981;99:1242-46.

## Diagnosing Amblyopia in Verbal Children

It is important to obtain a visual acuity as young as possible. With pediatric optotypes visual acuity can be measures as early as 2 years old. Be careful as to which pediatric optotype you use as many do not actually measure visual acuity which is ability to discern if two points are separated (e.g., two point discrimination.) Many pediatric optotypes have obvious shape and contrast clues that allow even a densely amblyopic child to see the figure. The Allen figures have been used for many years but they have significant shape and contrast clues so vision is overestimated. Your author has developed Wright pediatric optotypes (Copyright 2000) that greatly reduce shape clues as the all the figures are rectangular. Contrast clues are also reduced as the figures have equal blackness and parallel lines are avoided.

**Allen figures** seen right. Note how you can identify the "Tree" even when severely blurred (far right).

**Wright Figures** seen right. Note that they are not recognizable when blurred with a fogging lens (below).



**Wright Chart can be purchased:**

- wrightcenter2020@gmail.com

*Proceeds go to the Wright Foundation a 501c3 non-profit*

*Mocan MC, Najera-Covarrubias M, Wright KW. Comparison of visual acuity levels in pediatric patients with amblyopia using Wright Figures, Allen Optotypes, and Snellen Letters. J of AAPOS, Vol 9, N 1, Feb 2005, pp 48:52.*

## Amblyopic Vision

- **Amblyopia** is defined as 2 or more lines difference in best corrected visual acuity.

- ***Crowding Phenomenon:*** It is easier for amblyopic patients to see individual isolated optotypes than see optotypes in the middle of a linear display. To best identify amblyopia, test with a line of many optotypes rather than single optotype presentation. This is likely because amblyopic eyes have poor fixation.

- **Neutral Density Filter:** This filter reduces the transmission of light so it is analogous to turning down the lights. Reduced illumination reduces central vision more than peripheral vison. Since patients with dense amblyopia use extra-foveal retina, reducing illumination reduces the vision of the sound eye more than the amblyopic eye. So neutral density filters or scotopic conditions reduce the **difference** in vision between the sound eye and amblyopic eye in patients with dense amblyopia. Scotopic conditions does not, however, improve amblyopic vision.

**Question:** Visual acuity difference between the good eye and amblyopic eye is greatest when vision is tested with:

1. Patient looking through a neutral density filter
2. Single letter presentation
3. Optotypes that have shape clues like the tree in Allen Cards
4. A line of optotypes presented to the patient

**Answer:** Neutral density filters reduce light and create scotopic conditions that degrade vison of the good eye but not the amblyopic eye so the difference in vison is less with neutral density filters so # 1 is wrong. Single optotype presentation is easier for amblyopic eyes and underestimates amblyopia so # 2 is wrong. Optotypes with shape clues like the Allen tree are easy to see and underestimate amblyopia so # 3 is wrong. Better to use optotypes that accurately test acuity like the Wright Figures *(well duh)*. Optotypes presented in linear fashion is difficult for amblyopic eyes because of the crowding phenomenon and accentuate the difference between the amblyopic and sound eye. The answer is # 4 and linear presentation is best for detecting amblyopia.

## Treatment of Amblyopia

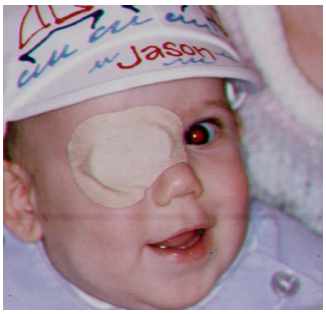
Early treatment is critical as delay can result in permanent visual loss. Treatment of amblyopia is based on two fundamental tenets:

1. Clear the retinal image
2. Force fixation to amblyopic eye if dominance is present by:
    - Patching
    - Penalization

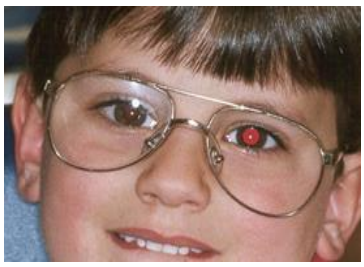
### Clear the Retinal Image:

The first stage in the treatment of amblyopia is to provide a clear retinal image. If there is a refractive error – correct it, if there is a media opacity – clear it. It makes no sense to patch the good eye if the image of the amblyopic eye is severely blurred. Amblyopia can often be treated by providing a clear retinal image without patching if the treatment is initiated early and there is no strabismus such as in the treatment of anisometropic amblyopia.

**Patching:** Patch the dominant eye to force fixation to the amblyopic eye. The end point is improvement in vision to within 2 lines of the good eye or 20/40 or improvement of fixation. Patients with fusion (i.e., anisometropia and ptosis) should have part-time occlusion to allow binocular fusion. Full-time occlusion is usually not needed but if used check the vision of the good eye at an interval of no more than 1 week per year of the child's age to check for reverse amblyopia of the good eye. For example, a 2 year old undergoing full-time occlusion needs to be examined every 2 weeks to make sure the vision good eye does not decrease.




**Penalization:** This is a treatment of amblyopia based on blurring the vision of the dominate eye by blurring the image with a fogging filter or pharmacologically with atropine. The idea is to blur the dominate eye and force fixation to the amblyopic eye. Atropine will not significantly blur the dominate eye unless the dominate eye is hyperopic. Photo right shows a child undergoing atropine penalization for a right amblyopia. Atropine drops are given once a day to the left eye and left spectacle lens is removed. The amblyopic right eye is given full optical correction. Atropine is no better than patching and not often used by the author.



## Treatment of Specific Types of Amblyopia

**Congenital Cataracts:** Early surgery is indicated in the first few weeks of life for monocular or binocular visually significant congenital cataracts. Postoperatively aphakic contact lenses should be placed within a few days after surgery. Spectacles are necessary for back up to the contact lenses. Initially monocular cataract patients should patch the good eye 50% of waking hours. Your loyal author published that very early surgery for monocular congenital cataracts first week of life can give excellent results and even stereo acuity.

Wright KW, Matsumoto E, Edelman PM. Binocular fusion and stereopsis associated with early surgery for monocular congenital cataracts. Arch Ophthalmol 1992:110(11);1607- 1609.

**Anisometropic Amblyopia:** Give the full correction first. If eyes are straight most will improve without patching or penalization. If after prescribing the full optical correction vision does not improve over 1 to 2 months then part-time patching of good eye is indicated. Penalization is an option, but it has not been shown to be superior to patching.

**Strabismic Amblyopia:** Patch the good eye to force fixation to amblyopic eye. Patch until the amblyopic eye can hold fixation well, or until vision has improved to within 2 lines of the good eye.

**Ametropic Amblyopia (Bilateral high hyperopia):** Bilateral high hyperopia $\geq$ +6.00 should be treated with full hyperopic correction regardless of the presence of strabismus even if orthotropic or exotropic. If the hyperopia is symmetrical then patching is not necessary. Do not cut the plus as amblyopic eyes do not fully accommodate and amblyopia will not improve.

**Question:** A 3 year old presents with orthotropia, has good stereo acuity and cycloplegic refraction of OD +1.00 sph, and OS +4.00 sph. You should immediately start full time patching of OD. True or False.
**Answer:** False- Most children with orthotropia and anisometropia are fusing and will improve with glasses alone. Full time patching may break down fusion. I would give OD +0.50 and OS +3.50 and if vision did not after improve after 2 months then start part time patching. Take 0.50 off both eyes to avoid blurring the good eye.

**Question:** A 5 year old presents with 20/80 OU and a cycloplegic refraction of + 8.00 OU. You should cut the plus and prescribe +4.00 OU – True or False?
**Answer:** False -Do not cut the plus. Bilateral high hyperopic children have bilateral amblyopia and will not accommodate for the uncorrected hyperopia- **Give the full plus!**

## Prognosis of Amblyopia

***The earlier the onset, the more distorted the image, and the longer the duration, the worse the prognosis. Early intervention is key to a good outcome.***

**Monocular Amblyopia:** In general strabismic and anisometropic amblyopia can be improved with treatment up to age 8 to 9 years. Rare cases of adults who go blind in their good eye will show improvement of vision of the amblyopic eye slowly over time. The presence of **eccentric fixation** indicates a poor prognosis with vision around 20/200 or worse. Eccentric fixation and dense amblyopia is commonly associated with late surgery after 2 months of age in patients with dense unilateral congenital cataracts.

**Bilateral Amblyopia**: Bilateral high hyperopia (> +6 .00 D) will cause bilateral amblyopia but prescribing the full plus even in older children will dramatically improve vision usually to 20/30 or better. The presence of sensory nystagmus associated with a bilateral lens or corneal opacity, however, indicates poor visual prognosis, usually around 20/100 or worse. One should not categorically give up on patients with sensory nystagmus and bilateral congenital cataracts as improvement can occur even in older children 8 to 14 years of age with visual acuity results as good as 20/60 (**Wright KW**, Christensen LE, Noguchi BA. Results of late surgery for presumed congenital cataracts. AJO 1992;114(4):409-415).

**Question:** A 6 month old has a partial cataract right eye. Which is the best sign that the cataract is amblyogenic and vision poor?

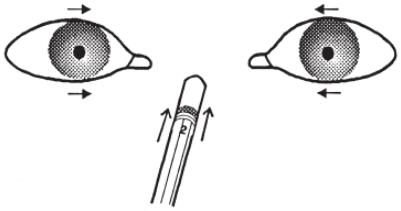
1. 3 mm in size
2. Orthotropia
3. Asymmetric red reflex
4. Inability to accurately follow a slowly moving target

**Answer:** A 3 mm cataract is not necessarily visually significant so # 1 is wrong. Orthotropia associated with a presumed congenital cataract is associated with a good visual prognosis (see my reference below). An asymmetric red reflex is an excellent screening test for a problem but does not indicate necessarily indicate poor vision, so # 3 is wrong. A better, objective measure of the opacity is to use a direct ophthalmoscope. If you can clearly see retinal detail then the opacity is not severe and very likely not amblyogenic. Answer # 4 is correct as poor fix and follow and the inability to accurately follow a slowly moving target indicates poor vision and amblyopia. If eccentric fixation is present this indicates poor vision and dense amblyopia. Remember diagnosing amblyopia in a preverbal child can be difficult. Use the entire clinical picture including history, fixation pattern, refractive error, density of opacity, and not just one factor to make your diagnosis.

# Chapter 6
# Accommodative and Acquired Esotropia

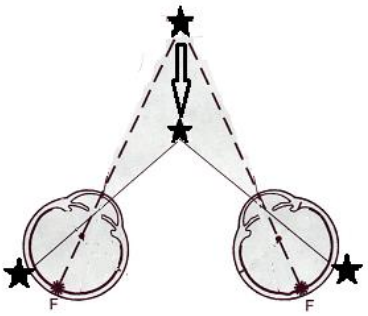
## **Near** Response

When an object moves towards us we invoke the near response*: accommodation, convergence, and pupillary constriction* to keep alignment on the target and increase depth of focus (drawing right).

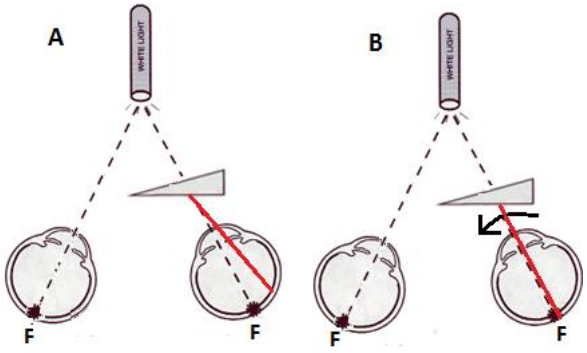


## Types of Convergence

There are three mechanisms that contribute to convergence.




- **Fusional Convergence:** Fusional convergence requires binocular fusion. As a target approaches, retinal images move to bi-temporal retina (see drawing right). Stimulation of bi-temporal retina results in a fusion reflex convergence to eliminate diplopia and align the eyes on the approaching target.




Note in drawings A & B (right) that a base out prism also moves the image to temporal retina and also induces fusional convergence. Measuring how much base out prism a patient can fuse is how we measure fusional convergence amplitudes. You can test convergence amplitudes at distance or near by using a base out prism bar. Increase the BO prism until fusion breaks which is known when convergence stops so the eye drifts out or the patient sees double. Most normal people can fuse 25 to 30 PD of BO prism.

### Normal Fusional Vergence Amplitudes

**Convergence = 25-30 PD**

**Divergence = 6-8 PD**

**Vertical vergence = 1-2 PD**

- **Accommodative Convergence:** This is convergence linked to accommodation. As the lens accommodates to focus on a near object there is a simultaneous reflex convergence. The relationship of amount of accommodation to amount of convergence is the basis for the AC/A ratio (see below). Accommodative convergence occurs without binocular fusion.

- **Proximal Convergence (Instrument Convergence):** When an object is near our eyes will naturally converge. This is convergence induced by the perception of an object being near is called instrument convergence as it occurs when viewing through an optical instrument such as a binocular microscope or an autorefractor.

## Measuring the AC/A Ratio

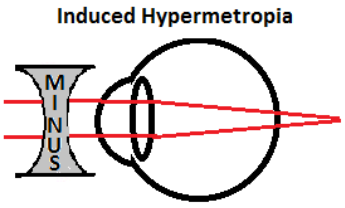
Accommodation and convergence are linked, as a certain amount of accommodation will produce a certain amount of convergence. The ratio of accommodative convergence to accommodation is termed the AC/A ratio. The AC/A ratio is measured by inducing a change in accommodation, then measuring the resultant change in convergence. Accommodation can be changed by using a minus lens to stimulate accommodation, or a plus lens to relax accommodation. Accommodation is also changed by changing the working distance; near induces accommodation and distance relaxes accommodation. . These two ways of changing accommodation are the basis for the two methods to calculate the AC/A ratio: 1) Lens Gradient Method (uses lenses) and 2) Heterophoria Method (uses near VS distance).

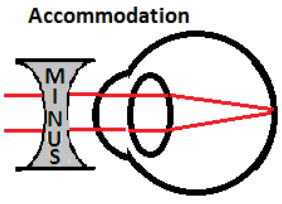
## Lens gradient method

The lens gradient method for determining the AC/A ratio changes accommodation by placing a lens plus or minus in front of the eyes, then measuring the resultant change in convergence:

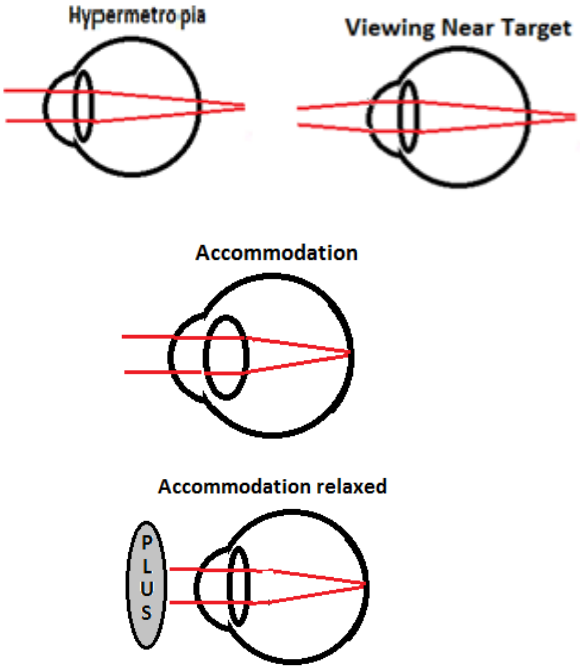
- Minus lens: Induces accommodation
- Plus lens: Relaxes accommodation

**Minus Lens:** A minus lens diverges light and pushes the image back inducing hyperopia (drawing below left). In response to the blurred retinal image the lens steepens to accommodate and bring the image in focus on the retina (drawing lower right). A minus lens induces accommodation which will result in increased convergence.



**Plus Lens:** A plus lens will converge light, thus relaxing accommodation needed for hyperopia and viewing a near target. The two drawings top right show the image is projected behind the retina with hyperopia and when viewing a near target. Drawing right middle shows accommodation needed to focus the image on the retina. Bottom drawing shows a plus lens in front of the eye to relax accommodation. Thus a plus lens reduces accommodation in patients who are hyperopic and for near viewing. Less accommodation results in less convergence.




## Lens gradient formula:

$$AC/A = \frac{\text{Dev org. - Dev w lens}}{\text{Lens power}}$$

- *Dev org* = original deviation (PD) measured without the inducing lens
- *Dev w lens* = deviation (PD) measured with an inducing lenses either plus or minus
- *Lens Power* (Denominator) = Inducing lens power in diopters

*For AC/A formulas exodeviations are minus and esodeviations are plus*

**Normal AC/A Ratio = 4 to 5 PD/D**

**Example:** A patient has a distance deviation of XT 20 without lenses. When -3.00 lenses are placed over both eyes the patient accommodates 3 diopters inducing convergence so the deviation now measures XT 5. The AC/A ratio is calculated below and is 5 PD/D:

AC/A = -20PD - (-5PD) /-3D

-20PD+5/ -3D

-15PD/-3D = **5 PD/D**

Note the lens gradient method is measured at a fixed distance and accommodation is changed by placing a plus or minus lens in front of the eyes. Because working distance is fixed, the inter-pupillary distance is not a factor, and not used in the formula.

**Question:** The 3 yr. old in the photos right has a cycloplegic refraction of +3.00 OU, and the following deviation:

Nsc ET 40 D (Photo right top)

Ncc+3.00 OU E 10 (Photo right bottom)

*Note: Nsc = near deviation without correction*

*Ncc = near deviation with correction*



Which of the following are true about the AC/A ratio?

1. Can't determine because you need the distance and near deviation to determine the AC/A ratio.
2. The AC/A ratio is normal 5 PD/D.
3. The AC/A ratio is high 10 PD/D.
4. Cannot determine AC/A ratio because the inter-pupillary distance is required.

**Answer:** The plus +3.00 lens reduces near accommodation by 3 diopters, which results in reduction of convergence by 30 PD as the esotropia changed from original deviation of 40 PD to 10 PD (i.e., ET40 - ET10). So for every diopter of accommodative relaxation there was a 10 prism diopter reduction of convergence so the AC/A ratio is 10PD/D.

AC/A = 40PD-10PD/3D = **10 PD/D - High AC/A ratio**

The lens gradient method is calculated using deviations measured at a fixed distance and can be distance or near so answer # 1 is wrong. The AC/A ratio is calculated to be 10 PD/D so answer # 2 AC/A = normal 5 PD/D is wrong. Answer # 3 is correct as the AC/A ratio is 10 PD/D which is high (normal $\leq$ 5 PD/D). Answer # 4 is wrong because the inter-pupillary distance is not used in the lens gradient method.

**Question:** At near patient has an XT 25 then you give -2.00 lens OU in glasses. If the AC/A ratio is 5 PD/D (normal) what do you expect the deviation is with the myopic correction?

1. ET 5
2. Orthophoria
3. XT 15
4. XT 30

**Answer:** The minus lens will stimulate accommodation thus cause convergence and reduce the XT. Since the AC/A ratio is 5 PD/D, there will be 5 prism diopters in reduction of XT for every diopter of myopia corrected. Minus 2.00 is given so the XT is reduced by 10 PD so the deviation without correction XT 25 goes to XT 15 with myopic correction and the answer # 3.

## Heterophoria Method:

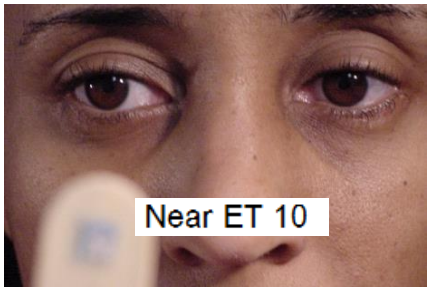
With the heterophoria method accommodation is changed by changing the working distance: comparing the deviation at distance VS near. Moving the target from optical infinity (≥20 feet) to 1/3 of a meter will induce 3 diopters of accommodation. The inter-pupillary (PD) distance is important for the heterophoria calculation because we are comparing alignment distance vs near. A patient with a wide PD will have to converge more for near than one with a narrow PD. Note this is often a board topic and remember the PD is in centimeters not millimeters.

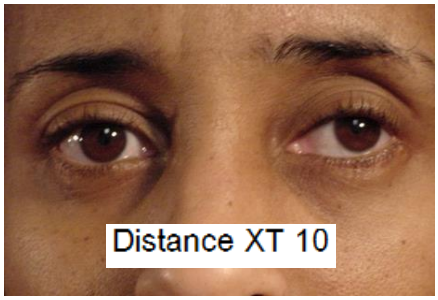
**Heterophoria formula:**

$$AC/A = PD_{(cm)} + \frac{N\ dev - D\ dev}{Acc\ change\ (D)}$$

- PD = inter-pupillary distance in **centimeters**
- N dev = near deviation in prism diopters
- D dev = distance deviation in prism diopters
- Acc change = accommodative change in diopters (D). Since no accommodation is needed for distance fixation the acc change is simply the near accommodation in diopters. Near accommodation in diopters is the reciprocal of the patient to target distance. If the fixation target is ½ of a meter from the patient the acc change is 2 diopters, and at 1/3 of a meter it is 3 diopters.

**Question:** Patient photos right has at near ET 10 and at distance an XT 10. PD = 50 mm
What is the AC/A ratio?




**Answer:** Use the Heterophoria formula-
5.0 cm + (20PD/2D)
5.0 cm + 10PD/D = **15 PD/D**

**Remember:** Accommodating to 50 cm is ½ meter and requires 2 diopters of accommodation. Also remember the PD needs to be expressed in cm, and 50 mm equals 5 cm. Don't forget that esotropia is positive and exotropia is negative.

*Clinically the Heterophoria method is virtually never used.*
*Also I hate the name it makes no sense- Hetero = Different, Phoria = latent strabismus.*

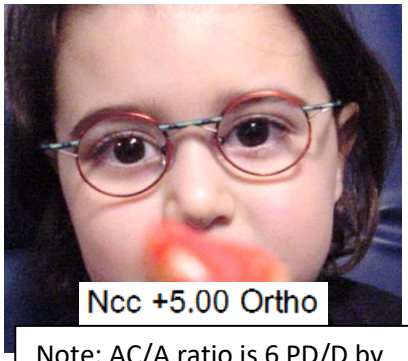
## Clinical AC/A Relationship

The late Dr. Marshall Parks used the clinical AC/A relationship to quantitate the distance-near differences of esotropia. Simply put, if an esotropia increases by more than 10 PD going from distance to near fixation then the AC/A relationship is high. This not a true AC/A ratio, but clinically it is very helpful. If on your oral boards someone asks you how you calculate the AC/A ratio don't freak out. Just say you prefer to use "Park's clinical AC/A relationship." If the esotropia increases by more than 10 PD for near the relationship is high.

- High AC/A Relationship = Dsc ET 20 PD and Nsc ET 35 PD, Difference 15 PD
  Dsc= distance deviation without correction, Nsc= Near deviation without correction

## Accommodative Esotropia

Accommodative esotropia is one of the most common types of strabismus. The onset ranges from infancy to late childhood with most becoming apparent around age 2 years. Usually the esotropia is intermittent and small at first then increases over time. The cause is usually high hyperopia but can be caused by a high AC/A ratio. Because of the high hyperopia the patient must invoke strong accommodation to see clearly. Increased accommodation causes increased convergence resulting is esotropia. The girl right has an accommodative esotropia (ET30) (upper photo). Her cycloplegic refraction is +5.00 OU and she must accommodate 5 diopters to focus which causes her to converge. The treatment is to give full hyperopic correction to relax accommodative convergence. When the full hyperopic correction is prescribed, accommodation relaxes and the eyes straighten to orthotropia (lower right photo). Because the eye straightens with hyperopic correction the treatment is to give full hyperopic correction, not surgery.


Note: AC/A ratio is 6 PD/D by lens gradient method (border line high)

The vast majority of patients with accommodative esotropia are hyperopic, usually greater than +2.00. Only occasionally will a patient be emmetropic and have a near esotropia due to a high AC/A ratio. Because accommodative esotropia is acquired and the eyes aligned during early visual development, the prognosis for recovery of binocular fusion and stereo acuity is relatively good. Hyperopia tends to diminish over time and about half of hyperopic children will grow out of their glasses in their teenage years.

## Characteristics of Accommodative Esotropia

- Onset acquired in infancy to 5 yrs. (2 yrs. average)
- Variable angle acquired esotropia
- Moderate to high hyperopia (+2.00 to +7.00)
- Relatively good binocular fusion potential

## Treatment of Accommodative Esotropia

Fundamental to treating accommodative esotropia is to prescribe the full hyperopic correction to reduce accommodative convergence and straighten the eyes. Hyperopia should be determined by a cycloplegic refraction. Cyclopentolate 1% is the standard, however if a patient is difficult to fully cycloplege then atropine 1% can be used. Refraction is performed 30 minutes to an hour after dosing for cyclopentolate and after 3 days of two times a day drops for atropine. In general try hyperopic correction in patients with acquired esotropia if the cycloplegic refraction is +2.00 or more. Patients should wear correction all waking hours. Most patients with accommodative esotropia require full hyperopic correction for many years some for a life time. Approximately 50% of patients will grow out of their hyperopia in their teen age years.

If the hyperopic optical correction straightens the eyes to within 10 PD of orthotropia for distance and near providing fusion no further treatment is needed. This is a pure accommodative esotropia and is correctable with single vision hyperopic spectacles. These patients have a good prognosis for obtaining binocular fusion and acuity as the esotropia is acquired so binocular fusion developed during infancy.

### Example of accommodative esotropia:

2 yr. old with acquired esotropia and cycloplegic refraction = +4.00 OU – glasses are given.

Nsc ET 20 → (+4.00 OU) → Ncc Orthotropia  Stereo acuity 50 sec

Dsc ET 20 → Dcc Orthotropia

*N= near deviation, D= distance deviation, sc= without, cc= with - correction*

**Note: Full hyperopic correction aligns the eyes and stereo acuity is achieved.**

**High AC/A Ratio Accommodative ET:** Some patients have a high AC/A ratio and after prescribing full hyperopic correction the alignment is corrected for distance but there remains a significant esotropia for near (ET > 10 to 15 PD). The treatment for these patients who are straight for distance but esotropic for near is bifocal glasses with a plus near add. If hyperopic (most are) give full hyperopic correction and a bifocal add usually a +3.00 (maximum near add). The goal of the bifocal add is to provide binocular fusion at near. The AC/A ratio slowly diminishes over time and most patients grow out of their bifocals in their teenage years.

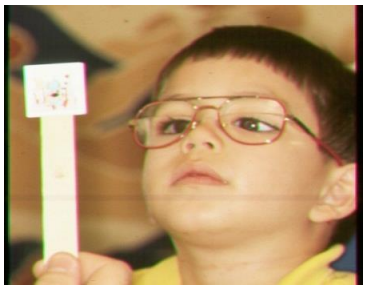
**Example High AC/A Ratio Bifocal Candidate:** 5 yr. old with acquired esotropia - cycloplegic refraction is +4.00 OU – glasses are given.

Nsc ET60 → +4.00 OU → Ncc ET 40 → ***+3.00 add*** → Ncc bif E 10, stereo=60 sec

Dsc ET 40 → Dcc Orthotropia

*D= distance deviation, N= near deviation, sc= without, cc= with - correction*

**Note: With +4.00 spectacles the patient is orthotropic for distance but there is a large esotropia for near. A +3.00 bifocal add corrects the near deviation to an esophoria with stereo acuity. NO SURGERY is needed - the treatment is bifocals.**



Photos right show the example patient with a high AC/A ratio wearing bifocals. Note when looking at near through the upper segment there is an ET 40 PD (upper photo), however, when the same near target is viewed through the bifocal add (lower photo) the eyes are aligned. These patients typically put their chin up to for near work and reading.

**Question:**

The 4 year old in photos above right is wearing bifocals with full hyperopic correction. He is orthotropia for distance through the upper segment. His near deviation is below:

Ncc upper segment = ET 30

Ncc +3.00 bifocal add = Orthotropia (fusing stereo acuity 50 sec arc)

*N= near deviation, cc= with correction*

1) Use the lens gradient method to determine the AC/A ratio.
2) Is this patient a surgical candidate?

**Answer:**

1. Lens Gradient AC/A = Change in near deviation with +3.00 lens:

Change in deviation with +3.00 PD = 30 PD

30 PD / 3.00 D = **10 PD/D** = high AC/A ratio

2. NO surgery because the patient is orthortopia for distance with hyperopic correction and orthotropia for near with bifocal add. Importantly patient is fusing with good stereo acuity.

## Miotics

There is a rarely used pharmacological treatment for high AC/A ratio accommodative esotropia and that is topical miotic drops such as phospholine iodide (i.e., echothiophate iodide). The treatment of choice is bifocal glasses, however, if the child does not tolerate the reading glasses then phospholine iodide drops once or twice a day is an option. Treatment is temporary as most children will grow out of the high AC/A ratio in their teenage years. Miotics, such as phospholine iodide are cholinesterase inhibitors and increase the effectiveness of locally released acetylcholine. Acetylcholine released in the ciliary body will last longer and produce more accommodation for a given amount of innervational stimulation. Thus, miotics reduce the accommodative effort to focus at near thus reducing accommodative convergence and reducing the esotropia at near. Miotics truly reduce the AC/A ratio.

**Adverse Effects of Miotics:** Phospholine iodide, even when given topically, is systemically absorbed and will lower cholinesterase activity in the blood for several weeks. Phospholine iodide prolongs the effect of the succinylcholine and may prolong respiratory paralysis after anesthesia. Succinylcholine anesthesia should be avoided if phospholine iodide has been used within 6 weeks before surgery. Systemic side effects of miotics may include brow ache, headaches, nausea, and abdominal cramping. If the lower dose of phospholine iodide is used, these complications are infrequent. Ocular side effects of phospholine iodide include iris cysts along the pupillary margin in 20% to 50% of cases, occurring at any time from several weeks to several months after treatment. Iris cysts tend to regress after discontinuing phospholine iodide. Phenylephrine used in combination with phospholine iodide may prevent iris cysts. Other rare and unusual complications include lens opacities, retinal detachment in adults, and angle-closure glaucoma.

**Partially Accommodative Esotropia:**

If a patient with accommodative esotropia still has a significant esotropia for distance after prescribing full hyperopic correction this is termed partially accommodative esotropia. The child in photo right is an example of partially accommodative esotropia as there is a residual esotropia of 25 PD for distance and near after wearing full hyperopic correction. This child has a constant esotropia and requires surgery is to establish binocular fusion. In general, surgery is indicated if there is a residual esotropia of 15 PD or more for distance. Bifocal glasses are not an option as they will not correct the distance esotropia.

## Surgery for Partially Accommodative Esotropia

Partially accommodative esotropia is a surgical disorder with the goal of establishing binocular fusion. Surgery is indicated for partial if after giving full hyperopic correction there is a residual esotropia of 15 PD or more for distance. Remember for board exams and in real life, it is important to repeat the cycloplegic refraction and give full hyperopic correction before surgery. **The surgery of choice is bilateral medial rectus recessions.** Early treatment gives the patient the best chance for obtaining binocular fusion.

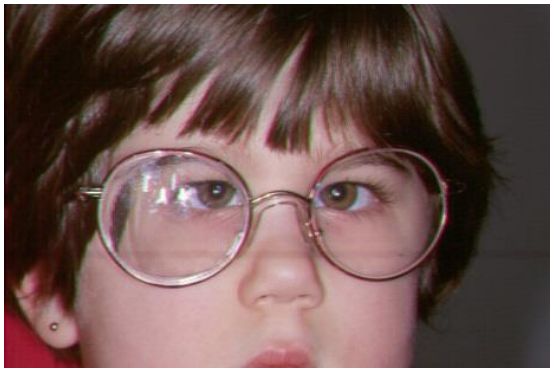
There are three approaches to determine the amount of surgery or target angle: 1. "Wright's Augmented Formula," 2"Standard Formula," and 3. "Prism Adaptation." **Wright's augmented formula** was developed by your author, and the target angle is an average of the near deviation without correction (largest), and distance deviation with correction (smallest). Successful outcomes are achieved in over 90% of cases operated using the Wright augmented formula and is my preferred formula (*see reference below*). The **standard formula** uses the distance deviation with correction (smaller angle) as the target angle. This formula has an unacceptably high incidence of undercorrections (25% -30%).

*Wright KW, Bruce-Lyle L. Augmented surgery for esotropia associated with high hyperopia. J Pediatr Ophthalmol Strabismus 1993;30:167-170.*

**Esotropia without correction**

**Esotropia with correction**

**Prism adaptation** determines the target angle by placing a base out (BO) press-on prism for the full deviation on the patient's glasses (photo right shows BO prism right eye). Then have the patient wear the glasses for a week to 2 weeks before re-measuring the deviation. If the deviation is then increases additional base out prism is applied. This process is repeated until the deviation is stabilized. This is less popular because of the extra time and expense of prism adaptation.



## Examples of Surgical Approaches to Partially Accommodative Esotropia

***1. Augmented Formula (Wright's)***

Nsc ET 40

Dcc ET 20

***Target angle = ET 30***

***2. Standard Formula***

Ncc ET 25

Dcc ET 20

***Target angle = ET 20***

*D= distance deviation, N= near deviation, sc= without, cc= with - correction*

***3. Prism Adaptation***

- Pre-adaptation = ET 30 so 30 BO prism applied to glasses
- After 2 weeks of wearing 30 PD prism deviation = ET 40 so 40 PD BO prism is applied to glasses
- After 2 weeks of wearing 40 PD the deviation has not changed so

  *Photo - Press on prism over OD*

  ***Target angle = ET 40***

**Question:** A 4 year old has a recent onset intermittent esotropia measuring 30 PD for distance and near. Cycloplegic refraction shows +4.00 D OU. What would you do?

1. Prescribe full hyperopic correction +4.00 OU.
2. Reduce plus so patient will tolerate glasses – give +2.00 D OU.
3. Surgery – bilateral medial rectus recessions
4. Bifocal glasses

**Answer:** Prescribe full hyperopic correction +4.00 OU so the answer is # 1. If the glasses correct the distance and near deviation then we are done – glasses is the treatment. If with the full hyperopic correction the eyes are aligned for distance but a residual esotropia is present for near > 10 to 15 PD then prescribe bifocal glasses with a +3.00 add. If there is a residual esotropia > 10 to 15 PD with full hyperopic correction for distance and near then strabismus surgery is indicated - bilateral medial rectus recessions. Wright's augmented surgery formula is preferred to determine target angle (*Wright surgery of course*.)

**Question:** 3 year old with an ET 20 PD distance and near after wearing +3.00 OU for 3 months. Multiple cycloplegic refractions verify a +3.00 sphere. Urgent surgery is indicated **True or False**?

**Answer:** True – Partially accommodative esotropia not fusing distance and near.

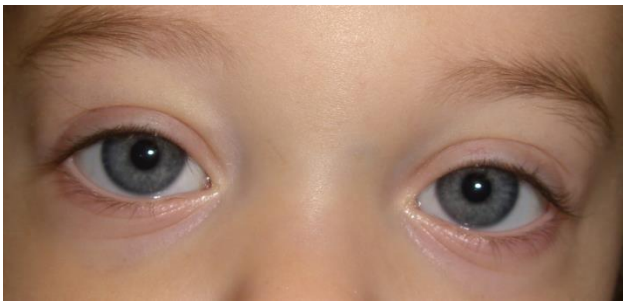
## Acquired Non-Accommodative Esotropia

**Children:** An acquired esotropia in childhood can occur without significant hyperopia (<2.00 D). In these cases it is important to rule out a neurological disorder. Check that ductions are full to look for a sixth nerve paresis. An important symptom that indicates a strabismus is acquired is the complaint of diplopia. Neurological disorders that can cause acquired esotropia include myasthenia, Arnold Chiari malformation, cavernous sinus tumor and hydrocephalus. In some neurological disorders such as Arnold Chiari malformation the esotropia may be comitant with full ductions. Photo right shows child who presented with a comitant esotropia and MRI showed significant *Arnold Chiari malformation* with large foramen magnum and herniation of brain. Neurosurgery resulted in improvement of esotropia but strabismus surgery was required. Unless there is an obvious cause for an acquired esotropia such as high hyperopia, consider obtaining a neurological evaluation including MRI brain. Treatment of acquired comitant esotropia is usually bilateral medical rectus recessions. These patients usually have binocular fusion potential so the target angle should be slightly larger than the angle measured.

**Adults:** A common type of acquired esotropia in older adults usually over 60 years old is divergence insufficiency esotropia. This is an intermittent esodeviation usually 10 to 20 prism diopters for distance and orthotropia for near. Patients present with diplopia for distance such as driving and watching TV which slowly becomes worse over many years. Neurological work up is usually indicated unless the duration of symptoms is long term and there are no signs of a sixth nerve palsy or other neurological signs. One theory on acquired esotropia in older adults is progressive lateral rectus muscle laxity and drooping of the middle of the lateral rectus muscles down, thus causing an esodeviation greater for the distance and often greater in downgaze. Initial treatment is often prism glasses especially for deviations < 10 prism diopters. Surgical treatment includes medial rectus recessions, tightening of the lateral rectus muscles (plication) or transposition surgery moving middle of the lateral recti up.

## Question:

The 3 year old patient in photo right has an acquired small angle intermittent esotropia of two months duration and associated with diplopia. Dolls head maneuver shows limited abduction left eye.

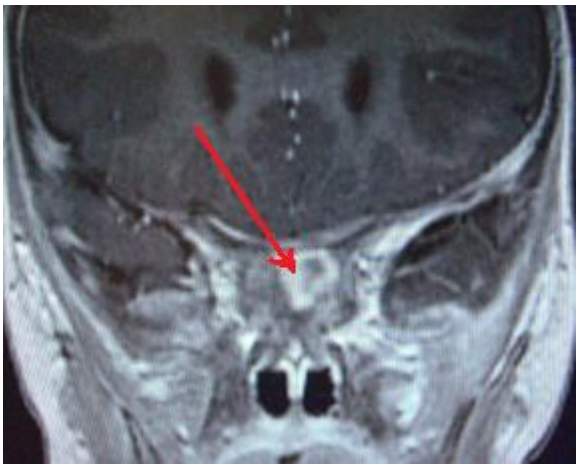


Cycloplegic refraction +1.00 OU
Fundus normal

Your management would be?

1. Prescribe glasses +1.00 OU then return in 6 months
2. Strabismus surgery to regain binocular fusion
3. Follow up in 6 months as strabismus often resolves
4. MRI of head

**Answer:** There are several red flags in this case! History of diplopia verifies the history that the esotropia is acquired. The patient is mildly hyperopic so may be accommodative esotropia but it's unusual that patients with accommodative esotropia have only +1.00 hyperopia. A key red flag is limited abduction. Accommodative esotropia should have full ductions. Acquired esotropia with diplopia and limited abduction might indicate an acquired 6$^{th}$ nerve palsy so a brain MRI brain scan is indicated to rule out a neurological etiology such as intracranial mass, or Arnold Chiari malformation. The answer is # 4.



Note: MRI was obtained and showed a tumor in the cavernous sinus (red arrow) consistent with a 6$^{th}$ nerve paresis and limitation of abduction left eye.

# Chapter 7 Congenital Esotropia

In general it is difficult to fuse esodeviations and much easier to fuse exodeviations. This is because our innate divergence amplitudes are weak measuring only 6 to 8 PD and our convergence amplitudes are strong measuring 25 to 30 PD. Weak divergence amplitudes explain why esodeviations tend to be constant tropias often associated with amblyopia and poor binocular vision, while exodeviations are well controlled and associated with high grade stereo acuity. Majority of newborns have a small exodeviation which is easily fused.

Congenital esotropia (AKA- infantile esotropia) is defined as a large angle esotropia that occurs in the first 6 months of life (photo right). The exact etiology of congenital esotropia is unknown. The deviation often increases over time. Abduction may be slightly restricted from tight medial rectus muscles, but the abduction saccade is intact. Alternating fixation indicates equal vision - no amblyopia and patching not required, while strong fixation preference means amblyopia of the deviated eye and patching of the dominant eye is indicated. Congenital esotropia is uncommon.

## Characteristics

- Onset: birth to six months
- Large angle (> 40 PD)
- Constant tropia
- Moderate hyperopia (+1.00 to +3.00)
- Poor prognosis for high grade stereo acuity
- Uncommon

## Differential Diagnosis

- Pseudoesotropia
- Infantile hyperopic accommodative esotropia (+3.00 or more)
- Congenital Fibrosis Syndrome
- Duane's Syndrome
- Mobius' Syndrome (sixth and seventh nerve palsy)
- Sixth Nerve Palsy - VERY RARE (hydrocephalus, infantile transient)

## Four Motor Associations

There are four characteristic abnormal eye movements associated with congenital esotropia listed below. Except for smooth pursuit asymmetry which occurs in early infancy, these motor abnormalities develop later, usually around 2 years of age. The top three of the four listed below probably represent the effects of disrupted binocular visual development as they are seen in other conditions that disrupt binocular visual development such as unilateral congenital cataracts. Since the acceptance of early correction of congenital esotropia promoted by your author (KW) these sequelae are now less common and less severe.

**Motor Associations of Congenital Esotropia**

- DVD or DHD (50 to 60%)
- Latent nystagmus (50%)
- Smooth pursuit asymmetry (100%)
- Inferior oblique overaction (60% - usually bilateral)

## Dissociated Strabismus Complex:

Early disruption of binocular fusion causes a dissociated type of strabismus that occurs relatively late after 2 years of age. There are three components: vertical (DVD), horizontal (DHD), and extorsion. In contrast to a true hypertropia where there is a corresponding hypotropia on the opposite side, DVD violates Hering's law as each eye drift up – thus the term "dissociated." In the photographs on the right note that with RE covered there is a right hyper and with LE covered there is a left hyper. Some patients will have mostly a horizontal deviation and this is termed dissociated horizontal deviation (DHD).

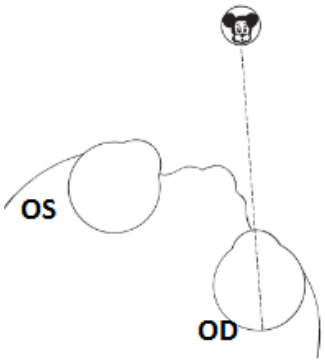
***Three components of DVD***

- Vertical – DVD
- Horizontal – DHD
- Extorsion

Treatment of DVD - If the DVD is a significant cosmetic issue surgery is indicated. It is usually bilateral so bilateral surgery is usually needed.

- Bilateral SR recessions or bilateral IR resections or plications
- If IO overaction and DVD coexists then IO anterior transposition is indicated (see Chapter 1 top of page 13 and Chapter 9 Oblique Dysfunction)

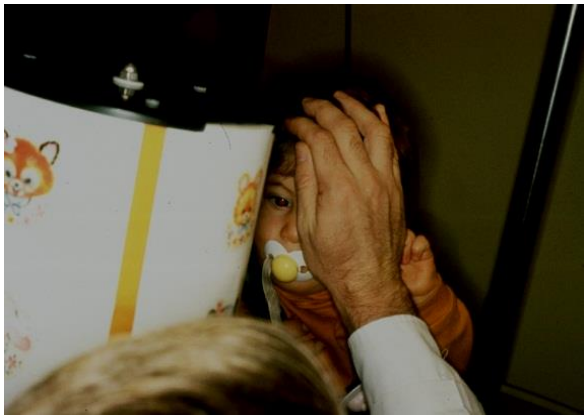
## Latent Nystagmus



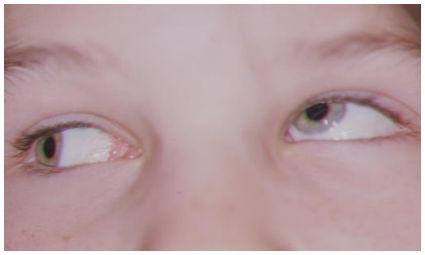

This is a horizontal nystagmus which is produced or worsened by covering one eye. It is characterized by the fast phase directed to the fixing eye. This means the fast beat goes to the right when the left eye is covered, and to the left when the right eye is covered. Patients can have manifest latent nystagmus (MLN) which means nystagmus is present to some degree even without covering one eye. The null point is in adduction so patients with MLN adopt a face turn so the fixating eye is in adduction. Drawing upper right shows the right eye fixing in adduction with face turn right. The companion photograph shows a patient with MLN fixing with the right eye in adduction to improve vision (null point), thus adopting a face turn to the right.

## Smooth Pursuit Asymmetry

Infants normally have smooth pursuit asymmetry with more accurate following when an object moves temporal to nasal. Pursuit is deficient and lags behind when an object moves nasal to temporal. This asymmetry is only seen when one eye is covered. With normal binocular visual development smooth pursuit asymmetry goes away and becomes symmetrical by 6 months of age. Disruption of binocular visual development such as occurs with congenital esotropia or a monocular congenital cataract causes permanent smooth pursuit asymmetry. This abnormality is life-long but does not interfere with everyday life as it only occurs under monocular conditions. The optokinetic drum (photograph right) is a good tool to demonstrate smooth pursuit asymmetry. Patients with smooth pursuit asymmetry will show poor or no OKN response when the drum rolls nasal to temporal and normal OKN response with rotation temporal to nasal. The presence of smooth pursuit asymmetry in an older child or adult indicates disruption of binocular visual development during infancy and is a marker for infantile onset of esotropia.



## Inferior Oblique Overaction (IOOA)

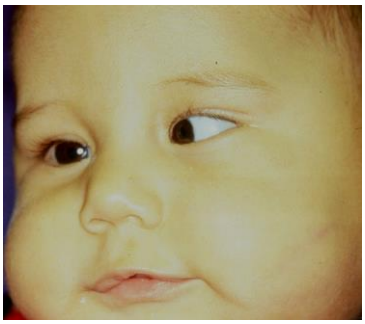


Inferior oblique overaction is characterized by:

- Up-shoot in adduction (photos right)
- V pattern - more divergence in up-gaze
- Usually bilateral

**Treatment:** IO weakening procedure - IO myectomy, or IO recession/graded anteriorization (see chapter 9)

## ET with Cross Fixation (Ciancia Syndrome)



Some patients with congenital esotropia have a large deviation (>60 PD) caused by very tight medial rectus muscles. Because the medial rectus muscles are tight both eyes are stuck in adduction and they adopt a face turn to fixate in adduction. If vision is equal they will cross fixate. Photo right shows cross fixation. The right eye is fixing in adduction with face turn right, then below the left eye is fixing in adduction with face turn left. These patients have restriction to abduction but because lateral rectus function is good there is a good abduction saccade. On attempted abduction there is exaggerated endpoint nystagmus as the patient struggles to abduct. Treatment is surgical – large bilateral medical rectus recessions (about 7 mm.)

**Ciancia Syndrome Triad**: 1) large angle esotropia, 2) face turn with fixing eye in adduction and 3) abduction nystagmus.

## Question:

A 40 year old presents with a history of previous strabismus surgery elsewhere for crossed eyes since birth. Your exam shows an alternating esotropia of 20 PD, nystagmus when one eye is occluded with fast phase to the fixing eye, R-hyper when RE covered and L-hyper when LE covered. Which one of the following is **unlikely**?

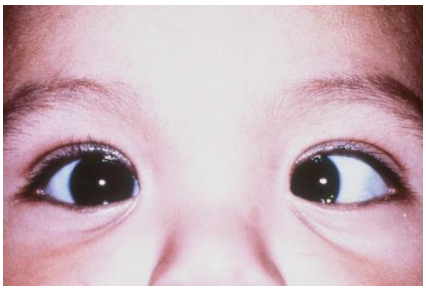
1. Bilateral DVD present
2. Patient has latent nystagmus
3. Monocular smooth pursuit asymmetry with poor pursuit nasal to temporal
4. Patient has amblyopia
5. Patient is stereo blind

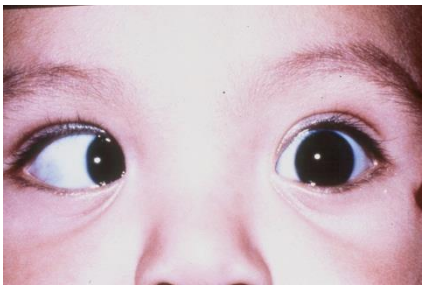
**Answer:** This patient has the typical motor signs of congenital esotropia, with bilateral DVD and latent nystagmus so # 1 and # 2 are true. The question did not specifically state the exam findings of smooth pursuit asymmetry but because of the evidence for congenital esotropia smooth pursuit asymmetry is likely so # 3 is true. The answer is # 4 as the patient does not have amblyopia because the patient has an alternating esotropia. Answer # 5 is true as the patient has a residual esotropia greater than 10 PD and that precludes binocular fusion so no stereo acuity.

## Treatment of Congenital Esotropia

In general strabismus surgery for congenital esotropia is bilateral medial rectus recessions. I prefer very early surgery at around 3 to 4 months of age if the baby is healthy (see why below), but for the board exam best to answer around 6 months of age. The exception to bilateral surgery is if the esotropia is associated with a blind eye, then perform surgery only on the blind eye. If the esotropia is associated with +3.00 D of hyperopia or more then glasses must be tried. If the hyperopic glasses straighten the eyes to within 10 PD of orthotropia then surgery is not indicated. A residual esotropia of greater than 10 to 15 PD after prescribing full hyperopic correction is an indication for surgery.

Photographs right shows a 6 month old with an alternating 70 PD ET with cycloplegic refraction +0.50 OU. There is no need for glasses or patching, so surgery is indicated.





**Preoperative Considerations – Full ophthalmic exam, plus:**

- Ductions – make sure full abduction or if slight restriction at least good abduction saccade (use dolls head maneuver)
- Versions – look for inferior oblique overaction
- Fixation Preference
    - Alternate fixation = no amblyopia
    - Strong fixation preference = amblyopia and need patching until child can hold fixation well with non-dominant eye
- Cycloplegic refraction (give full plus if $\geq$ +3.00)
- Fundus exam

**Surgical Goal** - Alignment to within 10 PD of orthotropia by 6 months to 1 year of age to obtain peripheral fusion (monofixation syndrome)

## Spontaneous Resolution of Congenital Esotropia

The NIH sponsored Congenital Esotropia Observational Study (CEOS) which yours truly KW was Study Chair, showed that spontaneous resolution of large angle (≥40 PD) congenital esotropia is rare. Small angle or intermittent esotropia, however, has a spontaneous resolution rate of approximately 30%. Small or intermittent esotropia in infancy should probably be followed for at least 10 months before considering surgery unless the deviation increases.

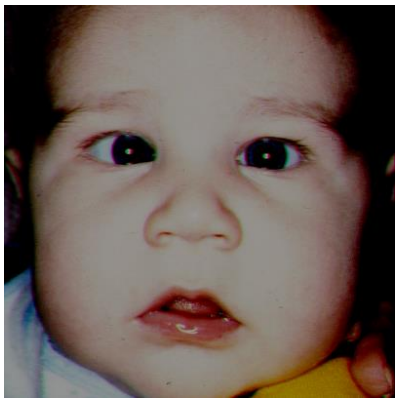
## Prognosis

Following standard dogma of performing surgery between 6 months to 1 year of age, approximately 50% to 70% will develop some peripheral fusion with low-grade stereo acuity (monofixation syndrome) if aligned to within 10 PD of orthotropia. With this approach, however, high grade stereo acuity is virtually *unobtainable*.

## Rational for Very Early Surgery


In the 1970's and 1980's Hubel and Wiesel Nobel Prize laureates, and Crawford and von Noorden clearly showed strabismus irreversibly disrupts early binocular cortical visual development. Using an animal model they demonstrated that even brief periods of esotropia in newborns caused permanent loss of binocular cortical neurons (figure right). Figure right shows that normally most of the occipital cortical cells are binocular "B" in figure A. Figures "B" and "C" show the central spike of binocular cells has diminished after only 18 or 20 days of prism induced esotropia. A multitude of basic science studies have verified these results. In 1994, I your loyal author reported clinical results of very early surgery operating at 3 to 4 months of age. 5/5 patients developed stereo acuity, with 2 achieving high grade stereo acuity of 40 seconds arc (*Wright KW, Edelman PM, McVey JH, Terry A, Lin M. High grade stereo acuity after early surgery for congenital esotropia. Arch Ophthalmol 199 4;112:913-919).* The combination of the animal studies and our clinical experience strongly suggests that very early surgery that obtains orthotropia can result in binocular fusion and stereo acuity.

*KW Note - Patients with intermittent strabismus who have straight eyes at times and have intermittent binocular fusion will maintain their fusion so in most cases they do not require urgent correction of the strabismus. It is the patient with a constant tropia and no fusion that require urgent treatment. Patients with congenital esotropia almost always present with a constant esotropia no fusion.*



## Question:

This 9 month old presents with an esotropia of 45 PD since birth. Patient alternates fixation, ductions are full and cycloplegic refraction is +1.00 OU. Which are most likely to be true?

1. DVD is present
2. Amblyopia present
3. Excellent prognosis for high grade stereo acuity.
4. Hyperopic glasses are indicated.
5. Esotropia will spontaneously resolve.
6. Surgery is indicated and sensory outcome is probably monofixation with peripheral fusion.

Answer: DVD develops later usually after 2 years of age so # 1 is wrong. Patient is alternating fixation so no amblyopia and #2 is wrong. High-grade stereo acuity is rarely achieved with congenital esotropia unless surgery is done before 6 months so # 2 is wrong. Glasses are indicated if $\geq$ + 3.00 so answer # 4 is wrong. It is rare that a large angle esotropia will resolve so # 5 is wrong. The correct answer is # 6 as monofixation is likely sensory outcome.

## Question:

All of the following are true regarding congenital esotropia except:

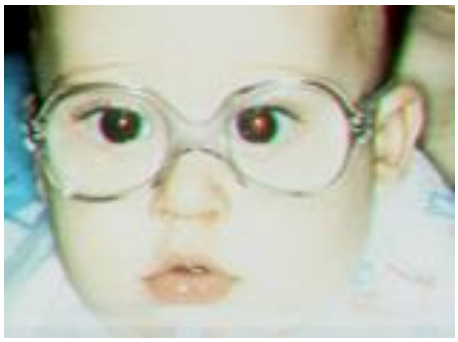
1. Preoperative patching the dominant eye is indicated if there is strong fixation preference for one eye.
2. Esotropia is common in normal newborn infants.
3. Normal reading ability even though smooth pursuit asymmetry is present.
4. Surgery should be performed around 6 months of age to obtain peripheral fusion.

**Answer:** # 1 is true as strong fixation preference indicates significant amblyopia. Newborns are often exotropic, but rarely esotropic so # 2 is wrong so the answer is # 2. Children with congenital esotropia have persistent smooth pursuit asymmetry with abnormal nasal to temporal pursuit; however, this is only seen when one eye is occluded. In normal life with both eyes open there is no pursuit asymmetry so reading is not affected so # 3 is true. Binocular fusion is obtainable with early surgery around 6 months of age so # 4 is true.

## Infantile Accommodative Esotropia

Infants 3 to 12 months of age can develop esotropia because they are hyperopic, usually +3.00 or more. This is called infantile accommodative esotropia and is in the differential of infantile esotropia. Because the esotropia is acquired they have a better prognosis than congenital esotropia and there is less chance that they develop DVD or latent nystagmus. The treatment is to prescribe the hyperopic correction based on a cycloplegic refraction. If the glasses align the eyes to within 10 PD of orthotropia, then glasses is the treatment and strabismus surgery in not needed. The baby in the photo above right has a cycloplegic refraction of +5.00 OU. Full hyperopic correction was given which resulted orthotropia so no surgery needed. If there is a residual esotropia of greater than 10 to 15 PD then this is termed partially accommodative esotropia and surgery is indicated. If amblyopia is present then treat before surgery. Surgery is bilateral medial rectus recessions with target angle for slightly more than the residual esotropia with glasses. Be sure to inform the parents that glasses will still be required after surgery.

## Question:



After wearing full hyperopic correction +5.50 OU for 8 weeks the infant in photo right (my son) has an ET 25 to 30 PD for distance and near with full ductions. We were patching right eye 2 hours a day and he now holds fixation well each eye.
***What did I do next?***

1. Obtain MRI scan of head
2. Increase patching right eye to 4 hours a day
3. Trial of bifocals
4. Urgent bilateral medial rectus recessions
5. Refer to an optometrist for vision therapy

*Note the bright red reflex from the deviated OS (positive Bruckner test)*

### Answer:

First answer is incorrect as patient has full ductions, and high hyperopia that partially corrects with glasses so the diagnosis is infantile onset partially accommodative esotropia and no MRI is needed. Answer # 2 is incorrect as previous patching has resulted in good fixation with patient holding fixation with either eye indicating that the amblyopia has resolved. Answer # 3 is incorrect because there is a significant esotropia distance and near- bifocals only works for near. BTW – We usually don't give bifocals to infants. Answer # 4 is correct and urgent surgery is what I did for the best chance to establish binocular fusion. Wait too long and fusion potential is lost forever. Answer 5 is ridiculous – no way.

**Note:** The patient shown above is my youngest of 5 children. I did the surgery myself – bilateral medial rectus recessions using my augmented surgery formula (see chapter 6) when he was 5 ½ months old. Thank goodness he did great! After wearing full hyperopic correction since infancy he grew out of his hyperopia in high school. Now 25 years after surgery he is orthotropic at distance and near and has perfect stereo acuity of 40 sec arc. Photo below shows my son and me at his graduation from the US Marine Corp boot camp. He was one of the best sharpshooters of the graduates – "chip off the old block!"

***BTW- No! I didn't operate on my son just to save money on the surgical fee –***

***I'm not that cheap!***

# Chapter 8 Exotropia

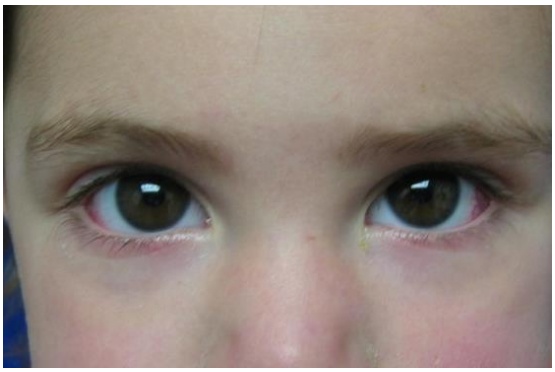
## Physiologic Exophoria

Our orbits are divergent so the normal eye position of rest is divergent, and small exophorias < 10 PD are considered normal. Innate fusional convergence is strong (25 – 30 PD) so fusing small exodeviations is easy. Over 70% of normal newborns have a small exodeviation which resolves within a few days to weeks..

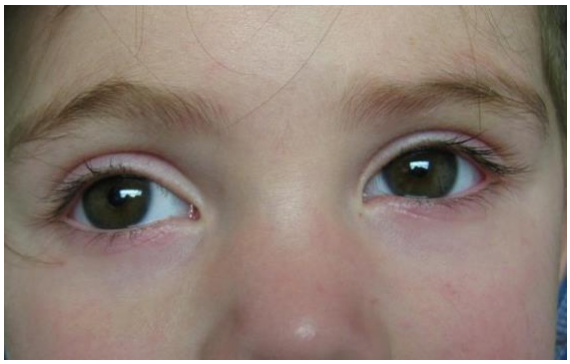
## Intermittent Exotropia

Intermittent exotropia is the presence of a large exophoria that can be fused because of our natural large fusional convergence amplitudes, but becomes a manifest exotropia at times especially when the patient is tired or day dreaming. Intermittent exotropia is by far the most common type of exotropia. The exotropia usually measures between 20 to 40 PD.

Photo right shows a child with intermittent exotropia and straight eyes because the deviation is fused by convergence (phoria phase) the stereo acuity normal 40 sec arc.

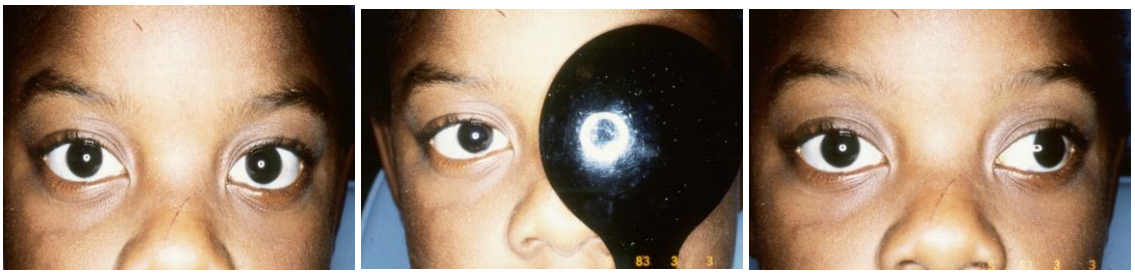


The right photo is taken moments later when the patient lost concentration so fusion broke and the exotropia has became manifest (tropia phase). When exotropic the patient has no stereo acuity and suppresses one eye.



Patients with intermittent exotropia have perfect stereo acuity (40 sec arc) when aligned (phoria phase), but no stereo acuity when tropic because the patients suppress the image from the deviated eye (tropia phase). Rarely patients will see double or have ARC when tropic. Often patients will squint to close the deviated eye (thus the nick name for strabismus “squint”). Typically intermittent exotropia (X(T)) becomes manifest when the patient is fatigued, daydreaming or takes a sedative. Alcohol is a depressant so imbibing an alcoholic beverage can bring out a latent exophoria. This is why the cowboy saw double after drinking in the saloon!

Covering one eye will break fusion and manifest an intermittent exotropia (see 3 photos below). Photo below left shows a patient with intermittent exotropia fusing with straight eyes. Middle photo shows covering left eye, and right photo shows the occluder removed and left exotropic. Prolonged alternate prism cover testing is used to dissociate the eyes and bring out the full exodeviation.



**Question:** Which of the following may cause the exotropia to become manifest in a patient with intermittent exotropia?

1. Covering one eye
2. Severely blurring vison of one eye
3. Taking a sedative like diazepam
4. Taking a few shots of whiskey
5. Attending a boring strabismus lecture by KW
6. All of the above

**Answer:** Covering one eye or severely blurring vision of one eye will break binocular fusion so answers one and two are correct. Answers three, and four are also correct because diazepam and alcohol are sedatives. # 5 is correct as one of my boring lectures makes one sleepy and will manifest a latent deviation. I can tell which resident has intermittent exotropia when I try to explain ARC – they immediately manifest their exotropia. So the answer is # 6, all of the above.

## Lateral Incomitance

It is well known that exodeviations are often incomitant with the exotropia decreasing in side gaze termed lateral incomitance. Older literature suggests reducing the amount of surgery if there is significant lateral incomitance, however, this practice has been mostly abandoned. The reason for the smaller exotropia in lateral gaze is probably due to the normal orbital end point of abduction. On lateral gaze the exodeviated eye will hit the lateral end point of abduction before the adducting eye thus collapsing the exotropia in lateral gaze.

## Key Points on Intermittent Exotropia

- Onset: usually after 2 years of age to teen age - or even occasionally adult onset
- Exodeviation can be fused but intermittently becomes manifest
- Excellent stereo acuity during phoria phase (40 sec arc)
- Suppression during tropia phase (diplopia or ARC rare)
- Do not have amblyopia

**Note:** Patients with intermittent exotropia do not get strabismic amblyopia because they have intermittent binocular fusion with high grade stereo acuity that provides binocular visual stimulation. Strabismic amblyopia occurs when there is a constant tropia and constant cortical suppression of one eye; such is the case in infantile esotropia with strong fixation preference for one eye. Patients with intermittent exotropia can have anisometropic amblyopia with the same incidence as the general population.

**Question:** Regarding intermittent exotropia which of the following is true?

1. Bifoveal fusion with high grade stereo acuity 40 sec arc
2. Cortical suppression
3. Amblyopia rare
4. Onset usually after 2 years of age
5. All of the above

**Answer:** All are true so # 5 is the answer. Remember when the eyes are aligned there is excellent binocular fusion and high grade stereo acuity but when the exotropia is manifest there is cortical suppression. Binocular fusion stimulates visual development of both eyes and prevents amblyopia.

## Treatment of Intermittent Exotropia

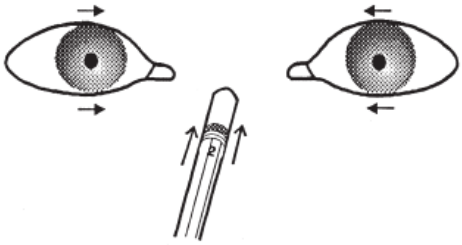
For the most part the treatment of intermittent exotropia is surgical. The indication for surgery is poor fusion control. Small to moderate exodeviations (< X 20 PD) that are well controlled do not need treatment. Most large deviations over 20 PD will eventually need surgery as they are difficult to fuse and the natural history is that the deviation usually increases over time. Non-surgical treatment can be used as a temporizing method for small angles but is rarely effective in the long term. One exception is convergence insufficiency as the preferred management is convergence exercises.

## Non-Surgical

In general non-surgical treatment is not effective except for the convergence exercises for convergence insufficiency. Below are non-surgical options.

- *Over minus glasses* - The idea is to reduce the exotropia by stimulating accommodative convergence by prescribing additional minus lenses over the refraction. Over minus is not well tolerated because it requires the patient to constantly over-accommodate. It is usually tried for small angle exotropia (10-15 PD) associated with concurrent myopia. Increase myopic correction by -2.00 to -3.00 over existing correction.

- *Convergence exercises* - Pencil push-ups or other convergence exercises can improve fusional convergence for near. It is useful for convergence insufficiency, but will not reduce the distance exodeviation.



- *Monocular occlusion* - Patching the dominant eye for 2 to 4 hours a day can in some children reduce the occurrence of the deviation. When the patching stops the deviation recurs. Monocular occlusion of the dominant eye works as anti-suppression therapy. If there is equal preference then alternate patch. I use monocular occlusion as a stalling method if the parents don't want surgery or if the child is too young for surgery.

## Surgical Indications

The most important indication for surgery is poor fusion control. If the deviation is difficult to control and becomes manifest more than 50% of waking hours then surgery is usually indicated. In general it is preferable to operate after 4 years of age. This is because a small consecutive esotropia often occurs after surgery and young children have the ability to suppress, so they can cause develop amblyopia and lose stereo acuity after surgery. Operate on children under 4 years old only if the exotropia is worsening and the deviation is not controlled. Older children with deviations greater than 30 PD are difficult to fuse and can cause eye strain so these patient should be considered for surgery.

## Surgical Plan

The procedure of choice for intermittent exotropia is **bilateral lateral rectus recessions**. Monocular recess/resect procedures induce incomitance and cause diplopia in side gaze. A small consecutive esotropia (E 4-8 PD) immediately after surgery is desirable as late recurrence of the exotropia is common. The consecutive esotropia will cause diplopia but will usually resolve in a few days. Patients should be warned that they may have post-operative diplopia. The standard surgical number charts have this small overcorrection built in. The pattern of the deviation is important for determining the surgical plan. Below is a classification based on difference of deviation distance vs near. See **Appendix I** of surgical numbers.

## Classification of X(T)

1. Basic X(T)
2. Convergence Insufficiency
3. Divergence Excess X(T)
   a. Pseudo
   b. True

### I. Basic X(T)

About 60% of intermittent exotropia patients have a similar deviation distance and near and are called Basic X(T).

Dsc X(T) 30

Nsc X(T) 35

TARGET ANGLE = XT 35  Bilateral LR recessions

### II. Convergence Insufficiency Exotropia

Patients with weak convergence have an exotropia greater for near. If the eyes are straight for distance it is best to avoid surgery and treat with convergence exercises (e.g., pencil push-ups). Convergence insufficiency is the one strabismus that can be helped by exercises.

Dsc Ortho

Nsc X(T) 30

CONVERGENCE EXERCISES – NO SURGERY

NOTE: If there is a significant XT (>15PD ) in the distance then consider bilateral lateral rectus recessions for about 5 PD more than the distance angle. Patients will usually require convergence exercises after surgery for an exotropia at near.

## III. Divergence Excess X(T)

Divergence excess is when the exotropia is larger for distance than near, by at least 10 PD. Divergence excess exotropia is the common pattern.

Near XT 15
Distance XT 30

There are two types of divergence excess intermittent exotropia:

- Pseudo - common (90%) - tenacious fusional convergence
- True - uncommon (10%) - High AC/A ratio

Tenacious fusional convergence is near convergence that persists for several minutes after monocular occlusion. Patients with pseudo-divergence pattern intermittent exotropia have strong tenacious fusional convergence that diminishes the near deviation. Patching one eye for 30 to 60 minutes will break tenacious fusional convergence. If the near exo-deviation increases after the patch test this indicates pseudo-divergence excess. If the near exodeviation does not increase with the patch test, this indicates true divergence excess and is associated with a high AC/A ratio.

**Note- Patients with divergence excess exotropia should have the patch test to determine pseudo VS true divergence excess.**

a. **Pseudo-Divergence Excess** – Patching one eye increases the near exodeviation to be similar to the distance deviation, and ok to operate for the full dissociated angle.

Dsc X(T) 30
Nsc X(T) 15 ➡ ***Patch test*** Nsc X(T) 30
TARGET ANGLE = 30 PD - Bilateral LR recessions

b. **True Divergence Excess** - If after the patch test the near deviation remains small the surgical target should be somewhere between the distance and near deviation around 20 to 25 PD in the case below.

Dsc X(T) 30
Nsc X(T) 15 ➡ ***Patch test*** Nsc X(T) 15
TARGET ANGLE = 20-25 PD - Bilateral LR recessions

NOTE: Patients with true divergence excess have a high AC/A ratio and may need bifocal glasses after surgery for an esotropia for near. True divergence excess can be a nightmare because of the high incidence of persistent esotropia and diplopia at near after surgery. Because of this tell patient that bifocals and more than one surgery is likely.

**Question:** 10 year old with intermittent exotropia manifest most of the time. Cycloplegic refraction is plano OU.

Dsc X(T) 40

Nsc X(T) 20

The most appropriate treatment plan:

1. Convergence exercises
2. Prescribe glasses -3.00 sph OU
3. Bilateral lateral rectus recessions for XT 40
4. Bilateral lateral rectus recessions for XT 20
5. Perform patch test to determine surgical target angle

**Answer:** Patient has divergence excess pattern with the distance deviation larger than near. The patch test is needed to determine the surgical target angle so the answer is # 5.

## Postoperative Management

A small initial postoperative over correction (ET 2-6 PD) is desirable, as this my reduce the chances for the exotropia to recur. Large consecutive esodeviations will often require further surgery such as seen in the photo on the right in this patient who is postoperative bilateral lateral rectus recessions.

A small consecutive esotropia can be initially observed as the esotropia usually resolves over a few days to a couple of weeks. Children under 4 years, however, can rapidly develop amblyopia so consider alternate patching 2-3 hours a day to prevent amblyopia if the esotropia is constant and persists for several days. For a persistent esotropia after a couple weeks consider prescribing base out prism glasses to eliminate the diplopia and preserve binocular fusion. Give just enough prism to allow fusion while leaving a small esophoria to build divergence. If after 4 to 6 weeks the esotropia persists then additional strabismus surgery should be considered. Either advance the lateral rectus muscles or recess the medial rectus muscles. Consider the possibility of a slipped lateral rectus muscle if abduction is limited and the esotropia is greater for distance. If the lateral has slipped resect the stretched scar and replace at the intended recession point.

**Question:** Your 15 year old patient has a poorly controlled intermittent exotropia.
Preoperative measurements:

Dsc X(T) 35

Nsc X(T) 10  Patch test = XT35

What do you want to do?

1. Patch for amblyopia
2. Prescribe bifocal spectacles
3. Bilateral lateral rectus recessions for XT 35
4. Bilateral lateral rectus recessions for XT 10

**Answer:** Intermittent exotropia is rarely associated with amblyopia so # 1 is false. After patch test the near deviation increased to the distance deviation so this is pseudo-divergence excess and surgery for the fully dissociated deviation is indicated (XT 35 PD). Note the exo-deviation is poorly controlled so surgery is indicated so the answer is #3.

**Question:** An 8 year old presents with a history of having surgery elsewhere for intermittent exotropia six months ago. The child has diplopia at near since the surgery. Your exam:

Ductions full

Dsc Orthotropia

Nsc ET 20

Which is most likely true regarding this case?

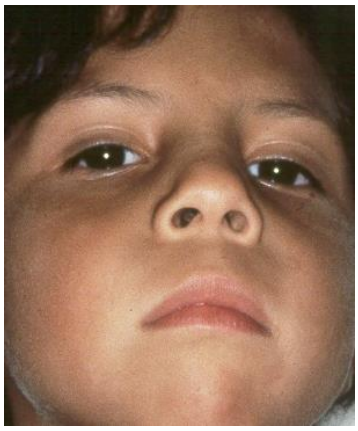
1. Patient has a high AC/A ratio.
2. Preoperative diagnosis was true divergence excess exotropia.
3. Bifocal glasses will help the diplopia
4. All the above

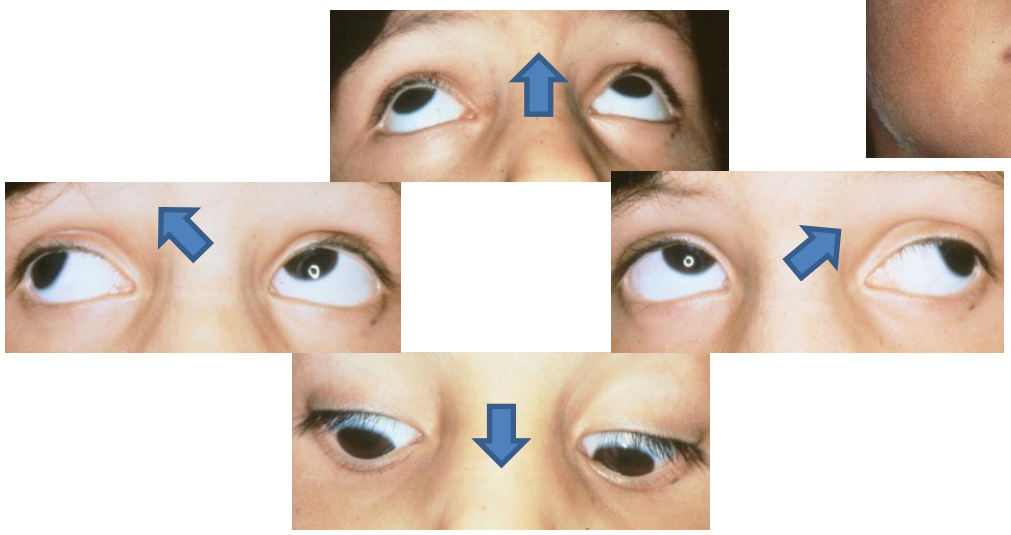
**Answer:** The patient most likely had a high AC/A ratio and true divergence excess exotropia prior to surgery as the eyes are straight for distance and there is a persistent esotropia for near so # 1 and # 2 are correct. The patient will benefit from bifocal add to correct the consecutive esotropia at near so # 3 is also correct, and the answer is all of the above # 4.

## Exotropia and Oblique dysfunction

### Inferior Oblique Overaction (Also see chapter 9)

Exotropia and inferior oblique overaction can coexist. Bilateral inferior oblique overaction gives a V pattern (greater XT in upgaze) so patients adopt a downgaze preference and chin elevation to avoid the large exotropia in upgaze. Patient in photo right has IO overaction OU and a V pattern so he adopts a compensatory chin elevation to aid in fusion. (He is not from Beverly Hills). The V pattern is associated with an up-shoot of the adducting eye on lateral gaze causing a LHT in right gaze and a RHT in left gaze (see composite photos below).

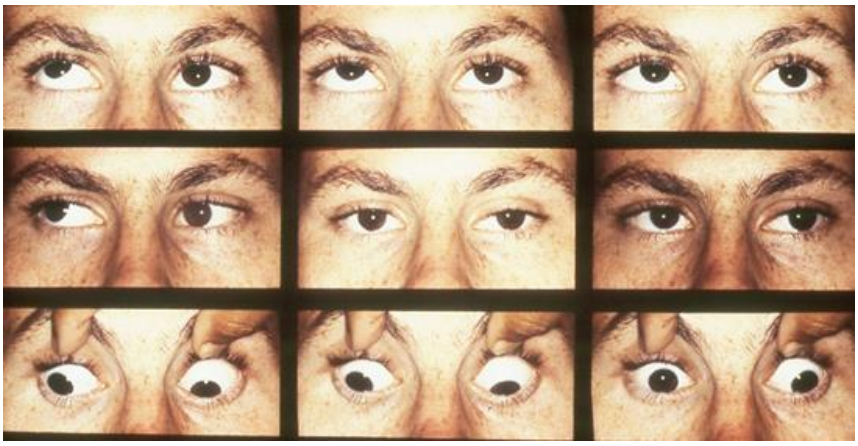




**Treatment:** If there is significant IO overaction and V pattern of 10 to 15 PD or more weaken the IO muscles at the time of the horizontal surgery. In general do not change the horizontal amount of surgery because of the IO muscle surgery. Inferior oblique muscle weakening procedures performed at the time of the exotropia surgery have good outcomes.

## Superior Oblique Overaction (Also see Chapter 9)

Bilateral superior oblique overaction is associated with an A pattern (increased XT in down gaze) and patients adopt a compensatory chin depression to keep the eyes in upgaze where the exotropia is diminished. The composite photos below show SO overaction left >right with an A pattern (Lambda subtype).



**Treatment:** In general try to avoid superior oblique weakening procedures in patients with intermittent exotropia as postoperative SO palsy frequently occurs causing cyclo-vertical diplopia. Instead displace the LR down to help correct the A pattern. If there is severe SO overaction and a large A pattern greater than 15 to 20 PD, then use a graded SO weakening procedure such as Wright SO silicone expander or a split tendon elongation (Chapter 9 P 123). A graded weakening procedure is preferred as it reduces the risk of consecutive superior oblique palsy.

## X – Pattern Exotropia

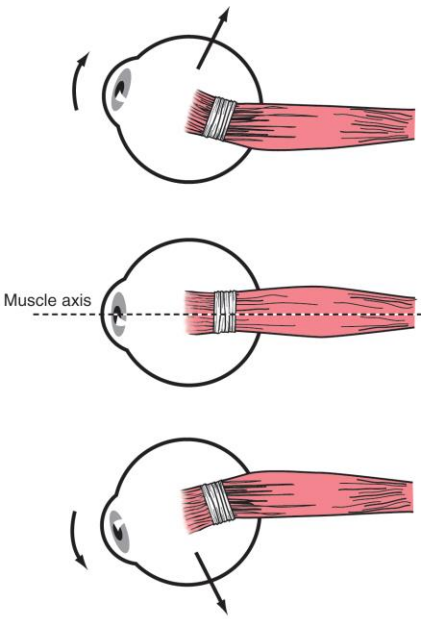
Patients with long standing large exotropia can have a X pattern with the exotropia increasing in up- and downgaze.

**Example:**

XT 30

XT 25

XT 30




The X Pattern is thought to be caused by tight lateral rectus muscles that flip the eye up and down on vertical gaze as shown in the drawing right. There is often associated mild pseudo IO and SO overaction. The X pattern disappears after bilateral lateral rectus muscle recessions.

## Question:

A 28 year old ophthalmology resident presents with poorly controlled intermittent exotropia and a chin elevation. Patching one eye eliminates the chin elevation. In addition to bilateral lateral rectus recessions which of the following is likely?

1. Need bilateral SO weakening procedure
2. Need IO muscle weakening OU
3. Need neck surgery
4. Chin up is because the patient is so proud that he/she understands the concept of ARC.

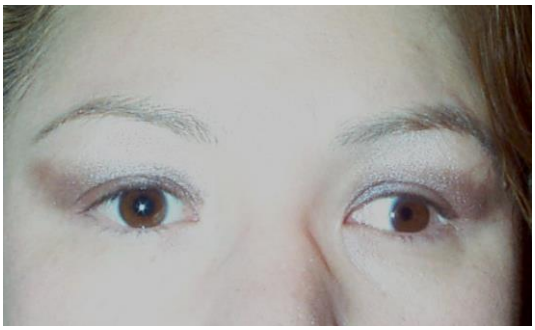
## Answer:

A chin elevation in a patient with exotropia is most commonly due to bilateral IO muscle overaction causing a V pattern. The patient adopts a chin elevation to move the eyes down where the exotropia diminishes. It is actually an inferior gaze preference manifesting as a chin elevation. Answer # 2 is correct as weakening the IO muscles is indicated for correcting the IO overaction and V pattern. Answer # 1 is wrong because SO overaction causes an A pattern and the gaze preference is up and chin is down. This is not a neck problem as patching one eye eliminates the chin posturing so # 3 is wrong. # 4 is also wrong because the patch eliminated the chin elevation thus the chin elevation is due to an issue with binocular fusion, not an attitude issue (haha).

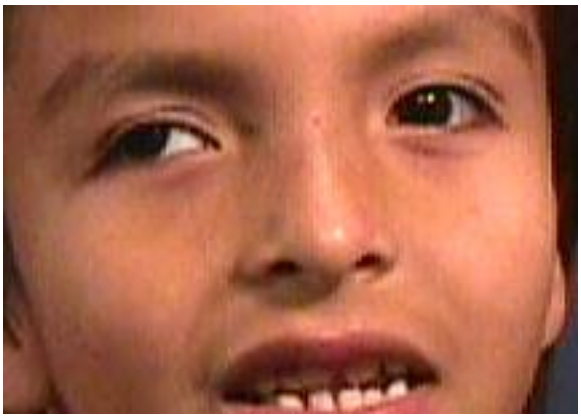
**Note:** Patching one eye is a useful way to determine if a face turn or head tilt is secondary to incomitant strabismus and the head posturing is to find a gaze position to obtain binocular fusion. If a patch eliminates face posturing then incomitant strabismus is the cause of the compensatory face posturing. Also a patch test can be used to determine if eye strain is caused by adifficulty obtaining binocular fusion, i.e. a phoria. If patching one eye makes the eye strain better, then the phoria is contributing to the eye strain.

## Sensory Exotropia

Sensory strabismus means the strabismus is caused by loss of vision and therefore loss of binocular fusion. Because our orbits are divergent our position of rest is divergent so a blind eye will often drift out which is called sensory exotropia (photo right). If blindness is congenital or occurs in infancy, esotropia may occur. The indication for surgery for sensory strabismus is based on cosmetic appearance. Surgery is isolated to the poor vision eye: either a recess – resect or recess – plicate procedure.



## Congenital Exotropia



Congenital exotropia is a large angle constant XT present at birth - with no fusion (photo right). It is extremely rare and most ophthalmologists will see only one or two cases during their career. Congenital exotropia is often associated with systemic disorders such as: prematurity, craniofacial anomalies, ocular albinism or cerebral palsy. The treatment for congenital exotropia is bilateral lateral rectus recessions, which should be performed after 6 months of age. There is a relatively poor prognosis for fusion similar to infantile esotropia. It has a much higher incidence of amblyopia than intermittent exotropia, with the incidence of amblyopia being similar to infantile esotropia (20 to 40%).

**Question:** Patients with which of the following conditions would you expect to have equal vision and excellent stereo acuity of 40 sec?

1. Infantile esotropia
2. Sensory exotropia
3. Intermittent exotropia
4. Congenital exotropia
5. None of the above

**Answer:** Infantile esotropia # 1 is wrong as onset is at birth or first few months the esotropia is constant and disrupts binocular fusion. Typically they have poor stereo acuity, monofixation syndrome, and have an incidence of amblyopia of 40%. Sensory exotropia is drifting because of a blind eye so they don't have stereo acuity so # 2 is wrong. Intermittent exotropia # 3 is correct as these patients can fuse the exotropia with innate convergence and have straight eyes much of the time, thus providing binocular stimulation. Patients have high grade stereo acuity 40 sec, equal vision, and intermittently become tropic especially when fatigued. Congenital exotropia like infantile esotropia is a constant onset at birth so it disrupts binocular development and associated with poor stereo acuity and amblyopia of 20% to 40% so # 4 is wrong.

# Chapter 9 A and V Patterns and Oblique Muscle Dysfunction

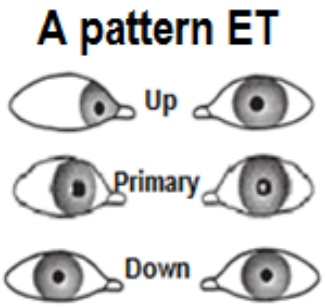
## A and V Patterns

A horizontal deviation (ET or XT) that changes when the patient looks up and down is termed an A or V pattern.

### *A pattern*

More convergence in upgaze and more divergence in downgaze by at least 10 PD. Drawing right show an A pattern esotropia.

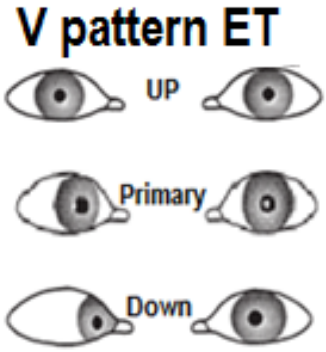
| A Pattern ET | A Pattern XT |
|---|---|
| ET 30 | XT 10 |
| ET 20 | XT 20 |
| ET 10 | XT30 |




### *V pattern*

More divergence in upgaze and more convergence in downgaze by at least 15 PD. Drawing right shows V pattern esotropia.

| V Pattern ET | V Pattern XT |
|---|---|
| ET 10 | XT 30 |
| ET 20 | XT 20 |
| ET 30 | XT 10 |



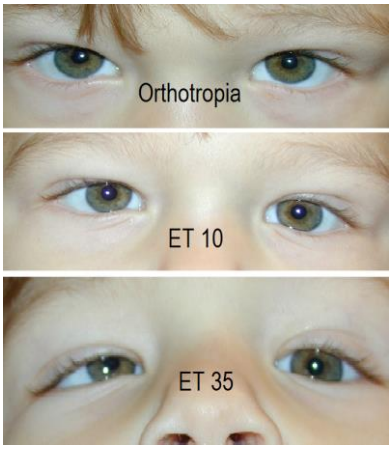

### Causes of A and V patterns

A and V patterns are often associated with oblique overaction or under action, but can be idiopathic without oblique muscle dysfunction.

**X pattern:** This less common pattern is usually seen in exotropia with increasing divergence in both up- and downgaze (see Chapter 8). It is probably caused by tight lateral recti and resolves after bilateral lateral rectus recessions.

## Question:

What pattern is seen in the photos right?




1. A pattern
2. V pattern
3. X pattern
4. No pattern

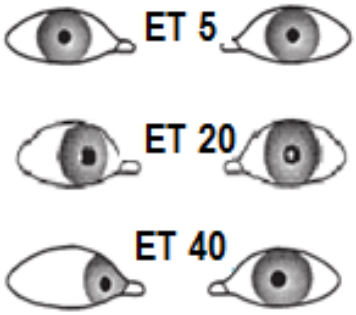
**Answer:** Orthotropia in upgaze, a small esotropia in primary position that increases in downgaze is a V pattern so # 2 is correct.

## Treatment of A and V patterns

If the pattern is small, not affecting fusion and not causing a compensatory head position then it can be observed. A significant pattern associated with oblique muscle dysfunction is treated by correcting the oblique muscle, such as inferior oblique weakening procedure for a V pattern associated with inferior oblique muscle overaction.


For the basic patterns without significant oblique dysfunction, you can transpose the horizontal recti in the manner shown in the drawing right. The medial recti are moved to the apex and the lateral recti are moved to the wide part of the letter. For a V pattern esotropia (no oblique dysfunction) perform bilateral medial rectus muscle recessions and move the new insertions down. In general move the muscles ½ tendon width (5 mm).

**Question:** Patient has an esotropia with the deviation shown right. There is no oblique dysfunction. What surgery is appropriate?

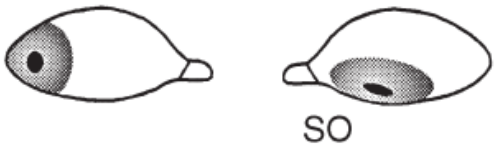



1. Bilateral medial rectus recessions with supraplacement
2. Bilateral medial rectus recessions with infraplacement
3. Bilateral lateral rectus resections with supraplacement
4. Bilateral medial rectus recessions without vertical transposition
5. Both 2 and 3

**Answer:** This is a V pattern esotropia. The answer is # 5 as both 2 and 3 are correct. One can either weaken the medial recti by recession or tighten the lateral recti with a resection or plication to treat the esotropia, and address the V pattern by transposing the medial recti to the apex (infraplace) and the lateral recti opposite to the wide part of the V (supraplace.)

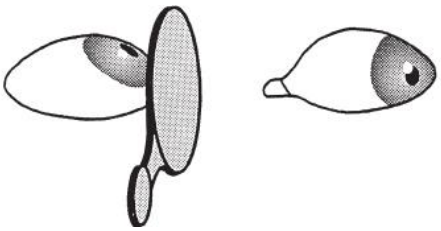

## Oblique Muscle Dysfunction

Oblique muscles can be dysfunctional and can over- or underact. Oblique muscle dysfunction can occur in isolation or associated with horizontal strabismus. Since there are 3 actions of oblique muscles, oblique over or under action will affect horizontal, vertical and torsional alignment in the field of action of the muscle. For example, superior oblique overaction produces a downshoot in adduction (see drawing upper right), excessive abduction in downgaze (A pattern), and intorsion worse in downgaze. Remember the field of action of the superior oblique is down and in, and inferior oblique is up and in.




Test for oblique muscle dysfunction by versions. Have the patient look in lateral gaze, and observe the adducting eye. It is helpful to cover the adducting eye with an occluder to break fusion to allow manifestation of oblique dysfunction. Hold the occluder at an angle so you can peak behind the occluder to see the adducting eye. Drawing right shows right inferior oblique overaction behind an occluder.

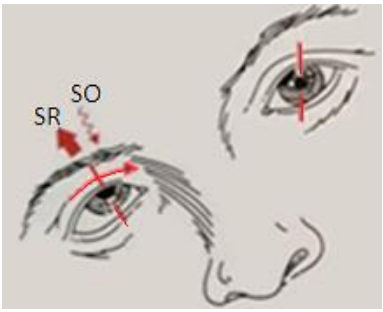
Oblique muscle dysfunction including oblique muscle palsy is a common cause of vertical strabismus. Think of possible oblique muscle dysfunction in patients with a hypertropia of otherwise unknown etiology.



## Bielschowsky's Head Tilt Test

The Bielschowsky head tilt is used to diagnose oblique and vertical rectus muscle palsies. It should be used on patients with a hypertropia of unknown etiology. No need for a head tilt test if the diagnosis has been made, such as in the case of a hypertropia secondary to thyroid related strabismus or a floor fracture. The Bielschowsky head tilt is most useful for diagnosing oblique muscle palsies specifically, superior oblique palsy. Clinically over 90% of significantly positive head tilt tests are associated with a superior oblique palsy. It is only useful for diagnosing an isolated single vertical or oblique muscle paresis.

Tilting the head to the side stimulates the vestibular leveling system of the eyes. Vertical rectus and oblique muscles have cyclo-vertical functions connected to the vestibular system and keep our eyes aligned when we tilt our head. A weak cyclo-vertical muscle will cause an imbalance of cyclo-vertical forces causing a vertical deviation that increases on tilting the head. Photos upper right show a positive head tilt with a right hypertropia greater on right head tilt in a patient with a right SO palsy. A positive head tilt test of 5 PD or more indicates a strong possibility that a vertical rectus or oblique muscle palsy is the cause of the vertical strabismus.




The drawing right shows the reason for the positive head tilt associated with a right superior oblique palsy. On tilt right the right eye intorts to keep the eye level. The SO and SR are responsible for intorsion so they are activated to intort, but there is a SO palsy. Since the SO (a depressor) is weak, the SR (an elevator) is strong and unopposed, so the SR pulls the eye up causing a right hypertropia on right tilt.

## Interpreting Head tilt

Dr. Marshal Parks described the classical three step test for diagnosing a cyclo-vertical muscle palsy. I have found a modification that I humbly call "***Wright's Rule***," a 2 step process, is simpler. ***Step 1:*** Do the head tilt test first. If the hypertropia increases on head tilt to the side of the hypertropia this indicates an oblique muscle palsy and if the hypertropia increases opposite to the hypertropia it is a rectus muscle palsy. ***Step 2:*** Test horizontal gaze to see where the hypertropia is greatest and match to the field of action of the cyclo-vertical muscle in question from step 1.

## Example of Oblique Muscle Palsy - *LHT in Primary*

Step 1. **Head Tilt** - LHT increasing on left tilt = an oblique palsy either:

Left SO palsy

or

Right IO palsy

Step 2. **Horizontal gaze**

If LHT increases in right gaze (field of action of the left SO) = L-SO palsy

If LHT increases in left gaze (field of action of the right IO) = R-IO palsy

## Example of Vertical Rectus Muscle Palsy – *LHT in Primary*

Step 1. **Head Tilt** - LHT increasing on right tilt = a vertical rectus palsy either:

Left IR palsy

or

Right SR palsy

Step 2. **Horizontal gaze**

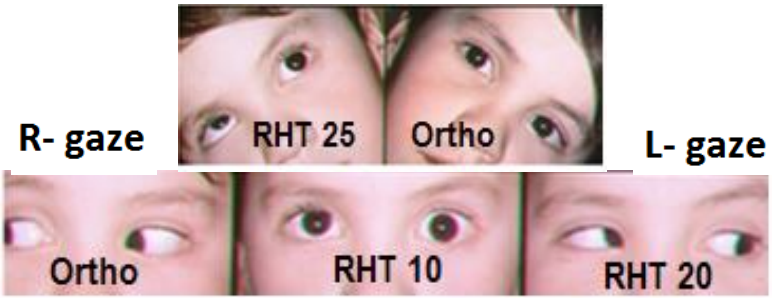
If LHT increases in right gaze (field of action of the right SR) = R-SR palsy

If LHT increases in left gaze (field of action of the left IR) = L-IR palsy

## Question:

Patient seen in the composite photos below has a RHT 10 in primary position. DIAGNOSIS?

1. Right SR palsy
2. Right IR palsy
3. Right SO palsy
4. Left SO palsy




***Answer:*** ***Step 1:*** Do the head tilt test first. Patient shows a positive head tilt with right hypertropia increasing on right tilt. This indicates an oblique muscle palsy so the rectus muscle answers (1 & 2) are wrong leaving answers 3 and 4 (right or left SO palsy). Since there is a RHT and a weak right SO muscle causes a RHT the answer is # 3 right SO palsy. Note: the head tilt test narrows the choice to an oblique muscle SO or IO. Since there is a RHT the weak muscle must be either the right SO (weak depressor) or a left IO (weak elevator). ***Step 2:*** The RHT increases in left gaze the field of action of the right SO (left gaze) so this indicates R-SO palsy.

**Note:** If you are confronted with a positive head tilt (i.e. board exam or clinical practice) and you forget the rules because you are too nervous and you have to guess – if there is a hypertropia that increases on head tilt - you will be right most of the time if you guess SO palsy on the side of the hypertropia.

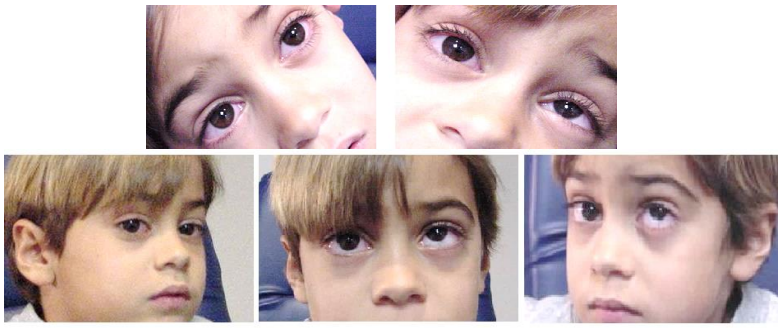
### *Question:*

Patient in photos right has a LHT 10 in primary that increases to LHT 25 in left gaze.

Head tilt: Right LHT 2, Left LHT 18

Which muscle is paretic?

1. Left IR muscle
2. Left SO muscle
3. Right IO muscle
4. Right SR muscle

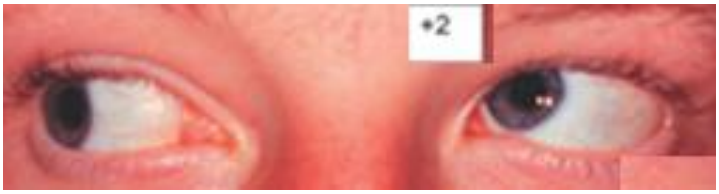


***Answer:*** *Do the head tilt first!*

Step 1. **Head Tilt** - LHT increasing on left tilt = oblique palsy either
Left SO (weak depressor)
Right IO (weak elevator)

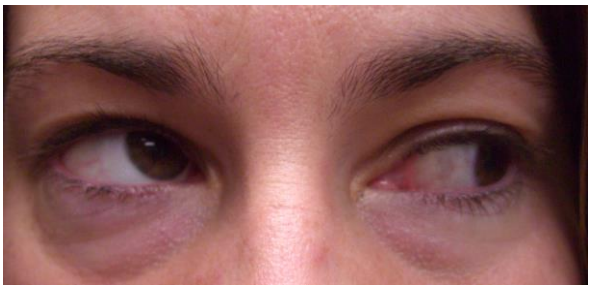
Step 2. **Horizontal gaze**
LHT that increases in left gaze (field of action of right IO) – R-IO palsy, answer # 3.
***Yes this is that rare case of isolated IO palsy, so be careful out there.***

## Inferior Oblique Overaction




Inferior oblique overaction (IOOA) is a common form of strabismus. It can be primary (idiopathic), or secondary caused by a superior oblique palsy (SOP). With primary IOOA head tilt is negative and with SOP head tilt is positive. The photo above shows a + 2 left IOOA with left up-shoot on rightgaze (dextroversion). Right eye is fixing allowing the adducting left eye to elevate. Bilateral IOOA is associated with a V pattern. The V pattern is because the inferior oblique muscles are abductors with the field of action in upgaze.

### *Primary IOOA*



Primary IOOA is usually bilateral but often asymmetrical. It is often associated with horizontal strabismus but it can occur on its own. Primary IO overaction is often associated with infantile esotropia (60%) developing around 2 to 3 years of age. Photo upper right shows the classical up-shoot right eye of IO overaction.

***Signs of IO overaction***:

- Up-shoot in adduction
- V pattern (Y subtype)
- Extorsion on fundus examination

The V pattern associated with primary IO overaction is a Y subtype with little change in the horizontal deviation from downgaze to primary gaze, then divergence from primary to upgaze. Photos right shows a patient with a V pattern esotropia. Note the eyes are straight in upgaze and there is an esotropia in primary and downgaze. This patient had an upgaze preference and adopted chin down posturing to obtain binocular fusion (upper right photo).

### *Secondary Inferior Oblique Overaction (Superior Oblique Palsy)*

Secondary IOOA is most commonly due to a unilateral congenital SOP. Since the SO muscle is a depressor a weak SO muscle can cause an ipsilateral IOOA and hypertropia in primary. A bilateral SOP will have canceling hypertropias so typically there is a small or no hypertropia in primary position. The key sign of SOP is hypertropia with the hypertropia increasing on tilt to the same side as the hypertropia, and hypertropia increasing on horizontal gaze opposite to the hypertropia (see above). To facilitate binocular fusion patients with a unilateral SOP will adopt a compensatory head tilt opposite to the side of the SOP, (e.g., R-SOP compensates with a tilt left). With primary IOOA the head tilt is negative. Extorsion can be seen on fundus exam. Patients with acquired SOP will experience torsional diplopia that can be measured on Maddox rod testing. However, patients with congenital SOP do not have subjective extorsion.

| | ***Unilateral SOP*** | ***Bilateral SOP*** |
|---|---|---|
| ■ Hyper in Primary | Large > 5 PD | Small < 5 PD |
| ■ "V" Pattern | Small < 10 PD | Large > 10 PD (Arrow Pat.) |
| ■ Maddox rod | Extorsion < 10 deg. | Extorsion > 10 deg. |
| ■ Head Tilt test | Hyper increases on tilt to the side of the palsy | RH tilt R and LH tilt L |

## Congenital SO Palsy

Congenital SO is usually unilateral with a relatively large hypertropia (10 -30 PD) which is intermittently fused so the eyes appear well aligned. Typically there is a compensatory head tilt and face turn away from the hypertropia to facilitate binocular fusion. Facial asymmetry is common with the smaller side dependent and opposite to the SO palsy. Photo right shows a teenage patient with a right congenital SOP. Note there is a head tilt left, face turn left (eyes right) and left face is smaller than right.

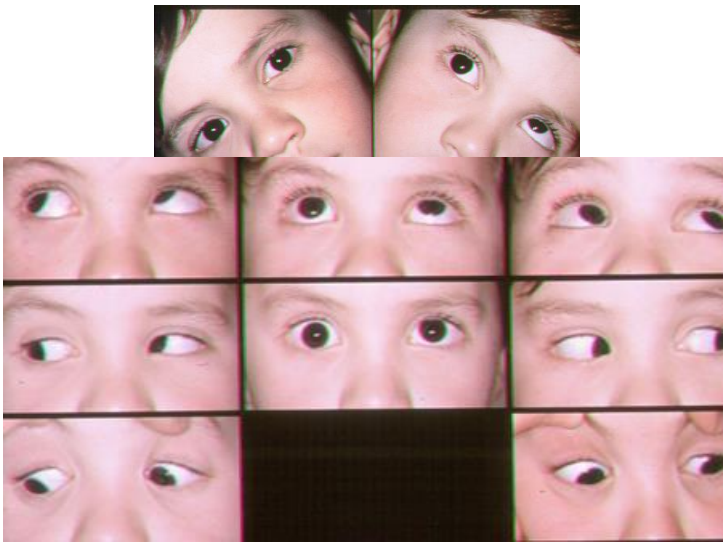
These patients have exaggerated vertical fusion amplitudes, and can fuse large hyper-deviations up to 25 to 30 PD, whereas normal vertical fusion amplitudes are puny at 2-3 PD. Patients usually have excellent stereo acuity. The hypertropia will become manifest when the patient is fatigued, much like patients with intermittent exotropia. When tropic, patients usually suppress diplopia but some will note double vision. There is usually significant IOOA, with minimal under action of the SO. The etiology of congenital SO palsy is unknown, but some cases have been associated with a lax or absent SO tendon (rare). Since patients have control over the deviation, they may present late even as senior adults. Check early childhood family photos to document longstanding head tilt.

**Note:** Often patients with congenital SO palsy are misdiagnosed and receive multiple consultations, CT, MRI scans, and even neck surgery for the torticollis! You can usually make the diagnosis on clinical grounds. In fact you can usually make the diagnosis as you enter the exam room. A patient with a head tilt, and facial asymmetry, and a long standing history of intermittent vertical strabismus has a congenital SOP until proven otherwise!

### Key Finding of Congenital SOP

- Large vertical fusion amplitudes
- Ipsilateral IO overaction
- Positive  head tilt  (> hyper on tilt to the hyper)
- Torticollis (compensatory head tilt opposite to SOP)
- Usually minimal or no diplopia
- No torsion on Maddox rod
- Facial asymmetry

Composite photo right shows left congenital SO palsy with large LHT in primary with the LHT increasing on head tilt left and in rightgaze ( L-IOOA).  Note the mild under action of the left SO muscle.



## Traumatic SO Palsy

Traumatic SO palsy is caused by closed head trauma.  It is almost always bilateral as the 4th nerves exit the brain close together so both nerves get traumatized with a traumatic shift of the tentorium.  Since the strabismus is acquired patients complain of torsional, vertical, and horizontal diplopia worse in downgaze (SO field of action).  Because both SO muscles are weak the verticals cancel each other so there is minimal to no hypertropia in primary position. These patients have significant torsional diplopia because there is bilateral extorsion which can be seen on fundus exam and on Maddox rod testing.  They see the fingers of the Snellen big "E" as slanting up because retinal images are inverted so extorsion is seen as slanting up. The pattern of strabismus for bilateral SO palsy is reversing hypers:  1) RHT on tilt right and LHT on tilt left, 2) RHT in leftgaze and LHT in rightgaze and a V pattern with esotropia in downgaze (see measurements below).

***Example Bilateral SO Palsy***

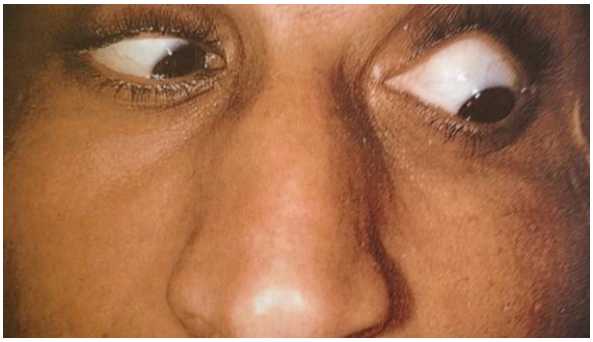
| Tilt R - RH 12 | | L - LH 10 |
|---|---|---|
| R- gaze | Ortho | L- gaze |
| LH 8 | RH 4 | RH 10 |
| | ET 10 | |

## Arrow Pattern of SO Palsy **(type of V pattern)**


Bilateral superior oblique palsy has a characteristic V pattern that is an arrow sub-type. Since the SO muscles are abductors in downgaze, bilateral SOP causes lack of abduction in down gaze thus causing an esotropia in downgaze. This causes a V because most of the eso-shift occurs from primary position to downgaze and is called an **arrow pattern (⇓)**. The composite photos right show big eso-shift from primary position to downgaze typical of an arrow pattern associated with bilateral SO palsy. The arrow pattern is virtually pathognomonic for bilateral SOP.

## Fallen Eye – Superior Oblique Palsy

"Fallen eye" occurs when the patients with SOP prefer to fixate with the paretic eye. Photo right shows a left fallen eye associated with a right SOP. The right (paretic) eye is fixing as it moves into its field of action - down and in. This movement requires great effort because the SO muscle is weak. Per Hering's law the yoke muscle the left inferior rectus also receives increased innervation but it is healthy, so it pulls the eye way down causing the "fallen eye." The large left hypotropia is the "fallen eye."



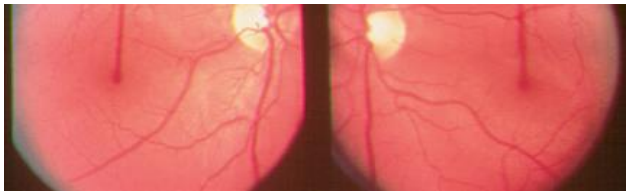
**"*Inhibition Palsy of the Contralateral Antagonist*"** is a complex and flummoxing term; however, it occasionally turns up on board exams. I will try to explain. It is really similar to the "fallen eye" as it occurs if the paretic eye is fixing. The yoke of the weak superior oblique muscle is the contralateral inferior rectus muscle. When the patient looks into the field of action of the weak SO (nasally), the SO muscle receives increased innervation to keep the eye down. Per Hering's law, the contralateral inferior rectus also receives increased innervation, and per Sherrington's law, it's antagonist the contralateral superior rectus muscle will receive increased inhibition. Increased inhibition of the contralateral superior rectus muscle gives the appearance that the contralateral superior rectus is paretic, thus the term "*Inhibition Palsy of the Contralateral Antagonist."* In the fallen eye photo above there is a right SOP but the left superior rectus muscle appears paretic as there is a large left hypotropia in levoversion.

## Masked bilateral superior oblique palsy

A bilateral asymmetrical superior oblique paresis can look like a unilateral superior oblique paresis as only one eye shows significant IOOA; this is termed *masked bilateral superior oblique paresis.* The presence of a V-pattern and bilateral extorsion on fundus examination also suggest bilateral involvement in patients with a presumed unilateral SOP. In these cases of masked bilateral superior oblique paresis, if surgery is performed on one eye, the contralateral superior oblique paresis will become evident postoperatively.

**Question:** The fundus photos lower right are most consistent with which of the following?

1. Bilateral SO palsy
2. Bilateral IO overaction
3. Masked bilateral SO palsy
4. All of the above

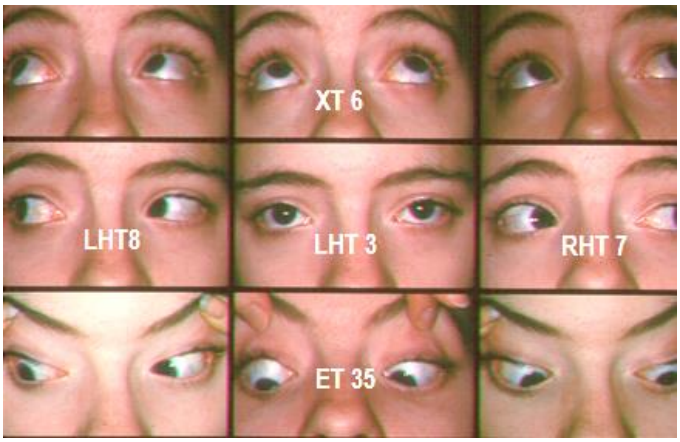


**Answer:** The fundus photos show bilateral extorsion with the foveas below the lower pole of the optic disc. The answer is # 4 as all are associated with bilateral fundus extorsion.

**Question:** The strabismus seen in the composite photos right has:
Head Tilt R- RHT 8, L- LHT10

This strabismus is most consistent with:

1. Bilateral inferior oblique palsy
2. Unilateral congenital SO palsy
3. Primary IO overaction
4. Traumatic bilateral SO palsy




**Answer:** Head tilt test is positive therefore most likely a SOP. There are also reversing hypers on head tilt: RHT on tilt right and LHT on tilt left, and lateral gaze = LHT in right gaze, RHT in left gaze with an arrow V pattern so this is classic for a bilateral SO palsy, answer # 4.

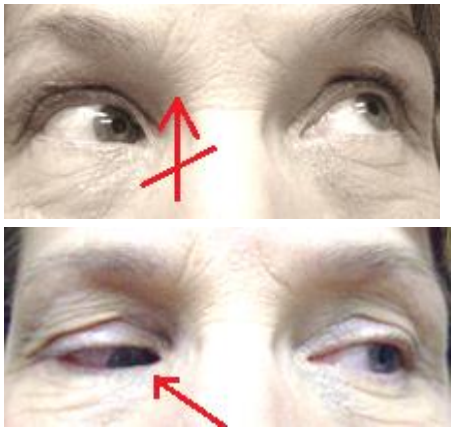
## *Primary Superior Oblique Overaction (SOOA)*

Primary superior oblique overaction is idiopathic and usually bilateral. As seen in the composite photos right the hallmark of SO overaction is downshoot in adduction and downgaze (arrows) and A pattern (lambda sub-type). Patients adopt a compensatory chin depression for binocular fusion as the eyes are best aligned with the eyes in upgaze. Fundus exam can show intorsion. Like IO overaction the head tilt test is negative. Onset is from birth but patients can present in adulthood. Patients usually have good binocular fusion and stereo acuity. Superior oblique overaction may be seen in esotropia, exotropia and occasionally without any horizontal strabismus. Antagonist inferior oblique function often is normal but there may be limited elevation in adduction, which is secondary to a contracted tight superior oblique muscle.

### *Lambda Pattern (type of A pattern)*

The superior oblique (SO) muscles are abductors with field of action in downgaze so overacting SO will cause divergence in downgaze producing an A pattern. The A pattern is actually a **lambda (λ)** subtype as most of the exo-shift occurs from primary positon to downgaze. See the composite photos right that show the lambda A pattern associated with superior oblique overaction (taken from composite above). Note that the exodeviation significantly increases from primary position to downgaze. This pattern is vertually diagnostic for bilateral SO overaction. Just like the arrow V pattent is diagnostic of bilateral SO palsy.

## *Inferior Oblique Palsy (Secondary SO Overaction)*

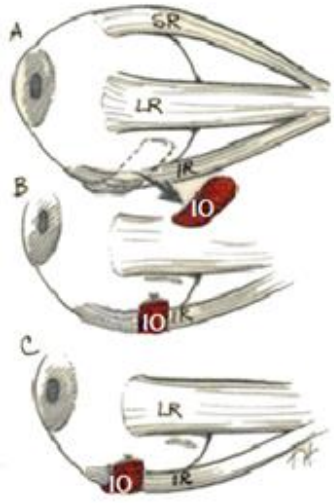


Inferior oblique palsy is rare and can be congenital or acquired. Congenital forms are idiopathic and almost always unilateral. Like congenital SO palsy, patients have large vertical fusion amplitudes and may present in adulthood. There is an ipsilateral hypotropia or contralateral hypertropia in primary that increases on tilt to the side of the hyper and on gaze to the side of the hyper. So a right IO palsy has a left hypertropia, that increases on head tilt left and in left gaze. Often patients have a compensatory head tilt opposite to the side of the hypertropia but ipsilateral to the IO palsy. Versions show limited elevation in adduction and secondary SO overaction with a down-shoot in adduction. Photos above right show a right IO paresis causing a left hypertropia that increases in left gaze, associated with right IO under action and right SO overaction. Limited elevation in adduction can look like Brown's syndrome, but in Brown's syndrome there is no SO overaction (Brown's syndrome see Chapter 11 P 150). A description of how to diagnose IO palsy with head tilt test is presented above, see Wright's rule and an example with composite photos are on page 113 in the question.

**Note:** Right upper figure (lower of the pair), shows right eye downshoot from SO overaction and **pseudo-ptosis**. The apparent ptosis is because the right eye is hypotropic and the lid follows down. Note that the pseudo-ptosis goes away when the right eye moves up (upper photo crossed out arrow).

## Surgery for Oblique Muscle Dysfunction

### Inferior Oblique Weakening




Inferior oblique overaction, both primary and secondary, can be treated by weakening the IO muscle. The three most frequently performed procedures to weaken the inferior oblique muscle include inferior oblique A) myectomy – remove a segment of muscle, B) recession – move insertion towards the origin to slacken the muscle, and C) anteriorization – move insertion anterior to equator to change the vector of forces so the IO is no longer an elevator – it is more or less vertically neutral. If the IO is placed anterior to the inferior rectus insertion then the IO will pull the front of the eye down and cause limited elevation. This is called **"anti-elevation."** It is a complication of placing the inferior oblique too anterior but can be used to treat DVD.

**Question:** The 8 year old patient had surgery for infantile esotropia at age 2 years. Now has strabismus seen in composite photos right and no diplopia.

Primary: Orthotropia
Head Tilt: R - Ortho, L – Ortho

1) Diagnosis?
    1. Bilateral SOP
    2. Bilateral primary IOOA
    3. Bilateral SR palsie
    4. Masked bilateral SOP

2) The most appropriate surgery:
    1. Bilateral inferior oblique recessions
    2. Bilateral inferior oblique myectomy
    3. Bilateral inferior oblique anteriorizations
    4. All of the above

**Answer:**

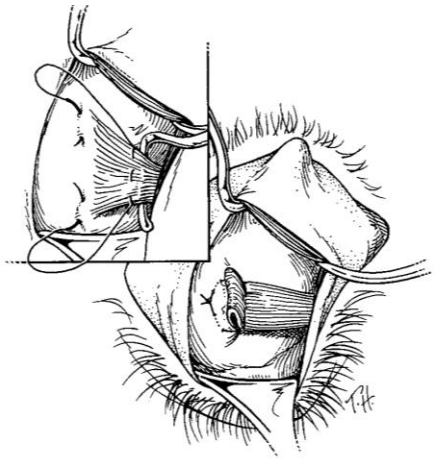
1) First off the head tilt test is negative so this is not a vertical rectus or oblique muscle palsy therefore answers # 1, # 3, and # 4 are wrong and by process of elimination the answer is # 2 bilateral primary IOOA. Note that the composite photos show bilateral IO overaction and V pattern. Actually the V pattern is a Y subtype because in primary the eyes are orthotropic and there is an XT in upgaze and this is indicative of primary IOOA.

2) A weakening procedure of the IO muscle is the treatment, either IO recession, myectomy or anteriorization so the correct answer is # 4. Anteriorization is often used by the author but avoid anti-elevation by keeping the IO muscle 2 mm posterior to the IR insertion and avoid the "J" deformity by keeping the IO posterior fibers posterior (see Chapter 1 P 13) .

## Superior Oblique Tightening

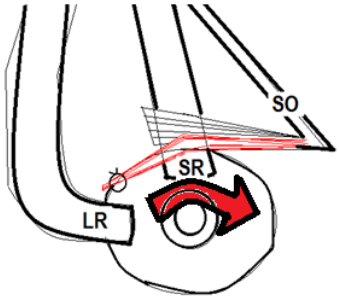
Two procedures that tighten the SO tendon include full tendon tuck and Harada Ito procedure which is a tightening of the anterior tendon fibers by a partial tuck or advancement of the anterior tendon fibers.

## Superior Oblique Tuck



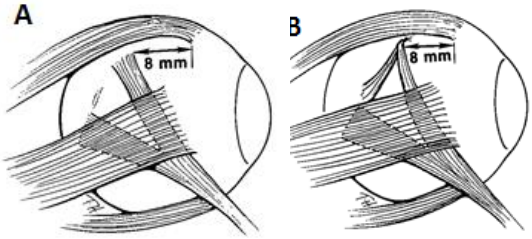
Extorsion associated with acquired SO palsy can be treated by tightening the anterior SO tendon fibers. The anterior SO tendon fibers are responsible for intorsion while the posterior fibers cause depression and abduction. The **SO full tendon tuck** (drawing right) tightens the tendon by pinching and folding the entire tendon. This results in intorsion, depression and abduction. It can be used in cases of a lax SO tendon causing a SOP. If the full tendon tuck is made too tight it will cause limited elevation worse in adduction due to tightening of the posterior tendon fibers, termed iatrogenic Brown's syndrome.

## Harada-Ito

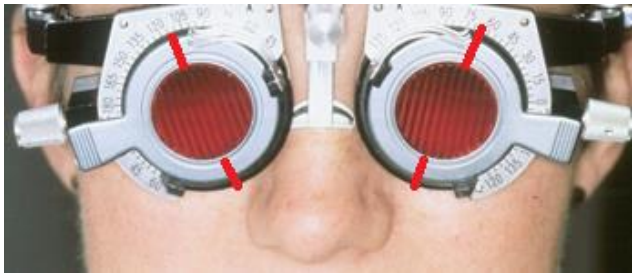



The **Harada-Ito** procedure specifically corrects extorsion as only the anterior tendon fibers are tightened so there is little depressor or abduction effect. Iatrogenic Brown's syndrome is uncommon. Harada Ito is the procedure of choice to correct extorsion without significant vertical strabismus. Drawing right shows the Harada-Ito as the anterior fibers in red are pulled temporally toward the lateral rectus muscle thus to intort the eye as shown by the red arrow. (Yes I did that drawing myself)

There are two ways to perform the Harada Ito: **A)** remove the anterior 1/3 of fiber and advance temporally towards the lateral rectus, or **B)** leave the tendon insertion intact but split the anterior fibers and pull them temporally towards the lateral rectus muscle (plication).




**Question:** Patient with a history of closed head trauma and torsional diplopia. Double Maddox rod is shown in photo right.



**Primary - Orthotropia**

**Head Tilt**; R- RHT 10, L- LHT 8

The most appropriate treatment is:

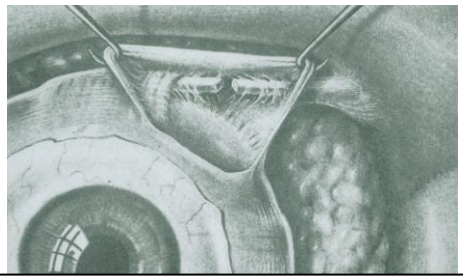
1. Prism glasses
2. Bilateral superior rectus recessions
3. Bilateral SO full tendon Tuck
4. Bilateral SO Harada Ito

**Answer:** The double Maddox rod shows bilateral extorsion. Most likely diagnosis is bilateral SO palsy causing extorsion. Prisms do not correct torsion so #1 is wrong. Harada Ito, # 4 is the best procedure as there is pure extorsion without a significant vertical deviation in primary position. A full tendon tuck is an option but can cause iatrogenic Brown's syndrome.

## Superior Oblique Weakening

Superior oblique weakening procedures can be used to treat superior oblique overaction or a tight superior oblique muscle tendon complex (i.e., Brown's Syndrome, see Ch. 10). The four most commonly used procedures include: tenotomy, tenectomy, Wright silicone tendon expander, and split tendon elongation. Beware of performing SO weakening procedures in patients with binocular fusion as post-operative SO palsy can result in intractable cyclo-vertical diplopia. Patients with intermittent exotropia have excellent binocular fusion, so if there is coexisting SO overaction it is better to infra-place the lateral rectus muscles than weaken the SO muscles unless the SO overaction is severe.

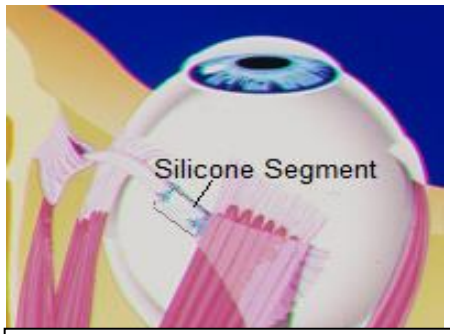
**Superior Oblique Tenotomy:** Berk described the superior oblique tenotomy as shown right to weaken the SO muscle. The tenotomy is performed nasal to the SR muscle. This is an effective procedure, however, it can result in SO palsy. Some including one of my mentors Dr. David Guyton advocates placing a suture bridge between the cut tendon ends to keep them from over-separating thus preventing a secondary SO palsy. This suture bridge is affectionately called a chicken stitch.



From Berk RN. Tenotomy of the superior oblique for hypertropia. Arch Ophthalmol 1947; 38:605.

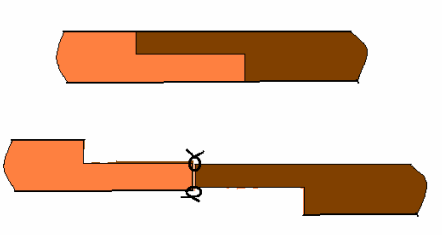
**Superior Oblique Tenectomy:** Is removal of a segment of SO tendon. This procedure has a high incidence of causing a secondary SO palsy and is not used often.

**Wright SO Silicone Tendon Expander:** The SO silicone tendon expander provides a graded slackening of the SO tendon. A 4 to 6 mm segment of a 240 or 40 silicone retina band is placed between the cut ends of the SO tendon to elongate the tendon (see drawing right). The incision is performed temporal to the SR muscle but the silicone segment is placed nasal to the superior rectus. Your author developed this procedure and is the procedure of choice in patients with binocular fusion to prevent a secondary SO palsy specifically for Brown's syndrome (Chapter 11).




Wright KW. Superior oblique silicone expander for Brown's syndrome and superior oblique overaction. J Pediatr Ophthalmol Strab 1991;28:101-107.

**SO Split Tendon Elongation:** The SO tendon can be elongated by splitting the tendon and uniting the split tendon ends as seen drawing right. In contrast to tenotomy, this will keep the integrity of the tendon intact. Practically it is somewhat difficult to perform and quantitate the elongation.



**Question:** Patient in composite photos right has intermittent exotropia and stereo acuity of 40 sec arc. Patient is bothered by the effort to fuse and wants strabismus surgery.

Dsc X(T) 30, Nsc X(T) 35

The preferred procedure is:

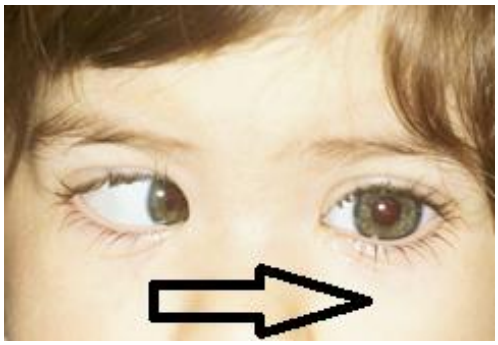
1. Bilateral LR recession
2. Bilateral LR recession and SO tenotomy
3. Bilateral LR recession with supra-placement
4. Bilateral LR recession and guarded SO weakening such as Wright SO tendon expander, SO tenotomy with chicken stitch, or split tendon elongation.

**Answer:** The composite photos were copied from P119 above and represents bilateral primary SO overaction associated with intermittent exotropia. Note the severe bilateral SO overaction with a large A pattern lambda subtype. Answer one is wrong as it does not address the severe SO overaction and large A pattern. Answer # 2 is wrong because this patient, as most patients with intermittent exotropia, has excellent binocular fusion and stereopsis. SO tenotomy has the high risk of inducing a SO palsy and intractable postoperative cyclo-vertical diplopia. Answer # 3 is wrong because for an A pattern the LR muscles are infra-placed. Answer # 4 is the best option as it addresses the XT, SO overaction and A pattern.

# Chapter 10 Restrictive and Paralytic Strabismus

Strabismus is easy – if an eye shows limited ductions there are only two causes:

- Restriction of ocular rotation
- Decreased muscle force



The photo right shows limitation of abduction left eye. It is important to diagnose why the left eye does not abduct. It could be restriction from a tight left medial rectus muscle or orbital obstruction, or paresis from a weak left lateral rectus. If you see an eye that has limited movement don't make the rookie mistake and call it restriction. You should just call it limitation until you determine if the cause is restriction or muscle paresis or both.

## Restriction of Ocular Rotation

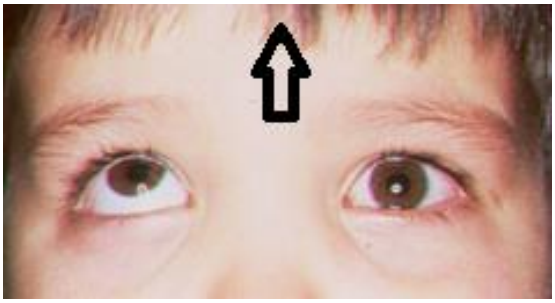
Restriction is a tethering of eye movement like a leash holding back ocular rotation. The cartoon right shows the classic description of restriction as being analogous to a dog on a leash. When the dog sees the cat by the tree the dog moves very fast and lunges for the cat. This fast movement is abruptly stopped by the restrictive leash. Likewise an important sign of ocular restriction is an eye that moves fast then stops abruptly.

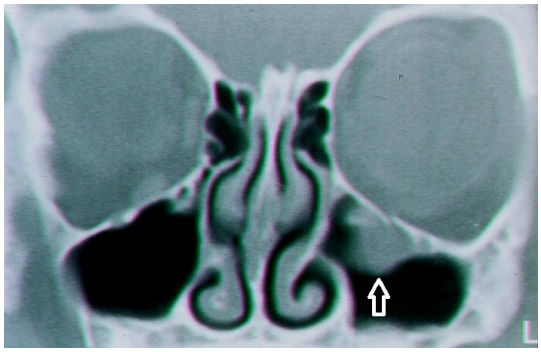
Ocular restriction has two causes:

- Tight muscle
- Peri-ocular adhesions to the globe

## Muscular Restriction (Tight Muscle)

A tight extraocular muscle will tether ocular rotation and cause restriction of eye movement. An example of muscle restriction is seen in photo right that shows limitation of elevation left eye. CT scan shows a left floor fracture with IR muscle entrapment that is restricting ocular elevation.

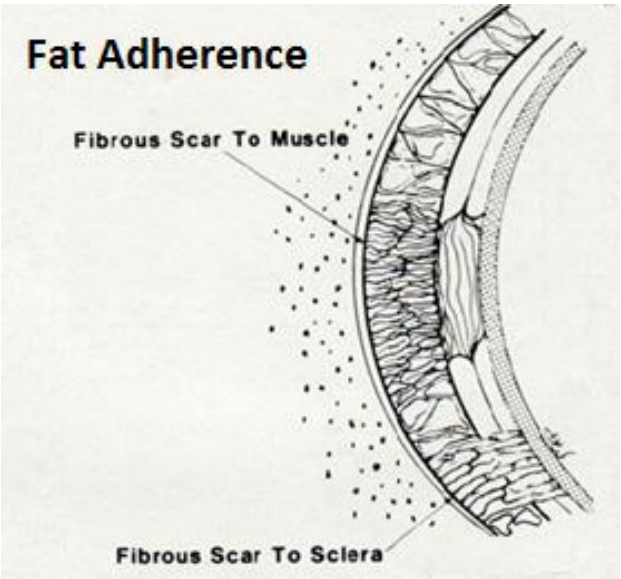




- Muscle entrapment orbit fracture
- Thyroid myopathy
- Congenital muscle fibrosis
- Secondary muscle contracture (antagonist of a long-standing paretic muscle)

## Periocular Adhesions Causing Restriction

If posterior Tenon's capsule is violated, adhesions to the extraocular muscles and/or globe can occur and cause restriction of ocular rotations. This is called fat adherence and is shown in the drawing right. Note scarring of orbital fat to extraocular muscle and the sclera. Fat adherence can occur after periocular surgery such as strabismus surgery or retinal detachment surgery. Normally the elastic Tenon's capsule surrounds the eye and separates orbital fat from the globe thus allowing free rotation with minimal resistance. Scarring of the conjunctiva such as in the case of recurrent pterygium can also cause restriction of eye movement.



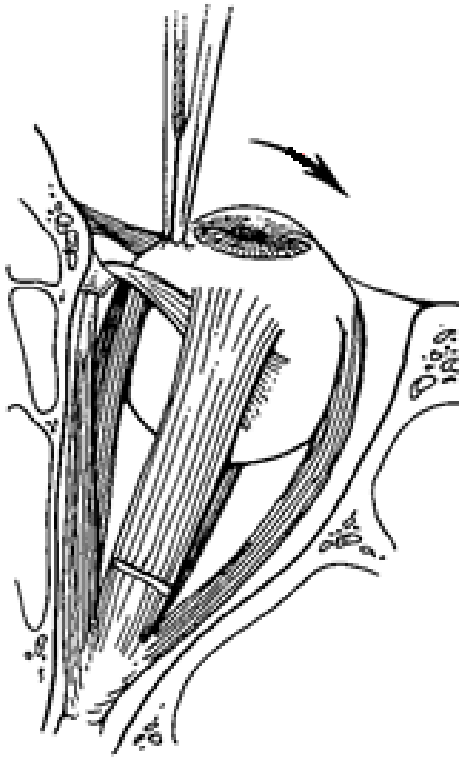
## Signs of Restriction

The two most important signs of restriction are:

- Dog on a leash motility pattern
- Positive forced ductions

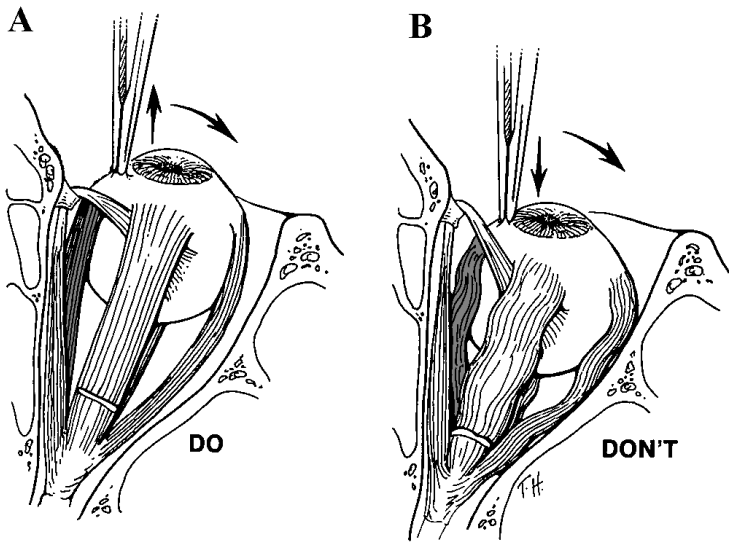
***Dog on a Leash –*** As described above (P 125), an important sign of restriction is the dog on a leash sign. If the cause of limited ductions is restriction the eye will move fast then stop abruptly when the eye hits the restrictive leash. Test for the dog on a leash sign by having the patient follow a fast moving target. Quickly move the target into the field of limitation and observe if the eye has a fast movement then stops abruptly. A fast eye movement that stops abruptly indicates a restriction.

***Positive Forced Ductions –*** The forced duction test consists of grasping the eye with a forceps and moving it to feel if there is restriction to ocular rotation. This is a passive test as the patient does not actively move the eye. Resistance to ocular rotation indicates restriction, and is termed positive forced ductions. The drawing right shows positive forced ductions from a nasal scar that limits abduction. Note the forceps are pulling the eye temporally but the fibrous band is restricting abduction. A negative forced duction test is when the eye can be rotated freely without significant resistance and rules out restriction. This test can be performed under general anesthesia or with topical anesthesia with patient awake. If awake, have the patient look toward the field of limitation.

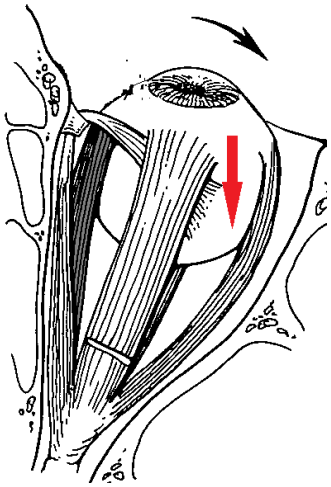


## Do's and Don'ts for Forced Duction Testing

When performing forced ductions to test for a possible tight rectus muscle be sure to proptose the eye to keep the rectus muscles on stretch (drawing A). If the globe is inadvertently retropulsed a tight rectus muscle might be missed as the rectus muscles will be slackened and forced ductions will feel free (drawing B).



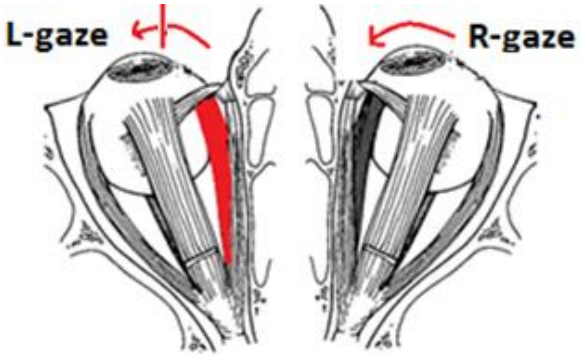

**Other Signs of Restriction**



- Lid fissure narrowing on attempted duction into the field of limitation indicates restriction. Drawing right shows a tight MR muscle and patient attempting to abduct against the tight MR muscle. As the LR pulls against the tight MR the eye is pulled posteriorly (red arrow).
- Increased IOP on attempted duction into the field of limitation indicates restriction. When the eye attempts rotation against a restriction the pull against the restriction increases the IOP. Test by taking the IOP with the eye turned away from the restriction (lower IOP), and repeat IOP measurement when the eye is attempting rotation into the field of restriction (higher IOP).

**Question:** If the left LR has good function and left MR muscle is tight all are true EXCEPT:

1. The left eye shows limited abduction.
2. There is an esotropia more in left gaze.
3. Left lid fissure narrows with increased IOP in left gaze.
4. Left eye shows slow abduction and no abduction saccade.




**Answer:** Drawing right shows levoversion in a patient with a tight left MR muscle (in red). The tight left MR muscle will restrict abduction of the left eye so # 1 is true. If the left eye can't fully abduct then on left gaze the right eye will over shoot to the left creating an esotropia more in left gaze so # 2 is true. On left gaze the left LR will pull the eye against the tight MR thus the eye will be pulled posteriorly causing lid fissure narrowing and the IOP will increase so # 3 is true. Answer # 4 is false and is the answer. Because the left LR has normal strength the eye can move fast with an abduction saccade until it stops abruptly when the eye hits the leash of the tight MR – i.e., dog on a leash (page 125).

## Rectus Muscle Paresis

Paresis of a rectus muscle will cause slowing and limitation of horizontal or vertical ductions in the field of action of the weak muscle. A lateral rectus muscle paresis will cause limited abduction. The term paresis means weakness while palsy indicates no muscle function.

### Causes of Rectus Muscle Paresis

- Cranial nerve palsy
- Congenital cranial nerve misdirection (Duane's syndrome)
- Lost or slipped muscle from strabismus surgery
- Direct muscle trauma
- Neuromuscular disease (myasthenia gravis, CPEO)

### Diagnosing Rectus Muscle Paresis

To diagnose rectus muscle paresis, one must access muscle function. The two most important tests of muscle function are:

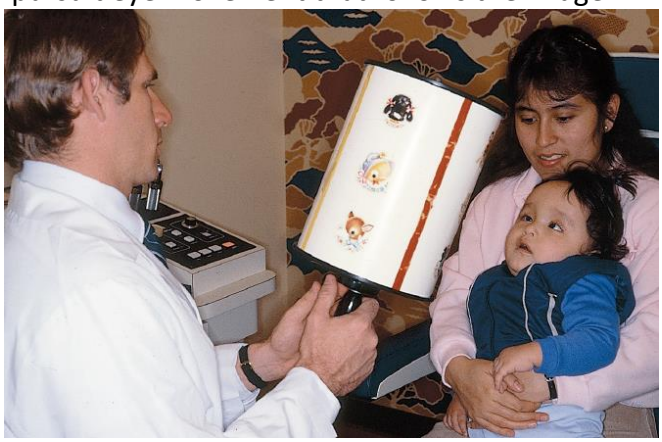
- Tests for saccadic movements
- Force generation test

***Tests for Saccadic Movements –*** Rectus muscles are responsible for generating saccadic eye movements (extremely fast). In order for a rectus muscle to generate a saccade the muscle must be at least at 70% of normal strength. If a muscle is weak it cannot generate a saccadic eye movement. Inability to generate a saccade is an important sign of rectus muscle paresis. Even a small amplitude fast eye movement indicates good rectus muscle function.

## Tests for Saccades

**Fast versions:** You can test for saccadic eye movements by simultaneously presenting two fixation targets on the patient's right and left for horizontal movements, and up and down for vertical movements. Have the patient quickly alternate fixation between the two fixation targets and observe for fast eye movements. Photo right show testing of horizontal saccades with patient directed to quickly look at the left then right finger back and forth.

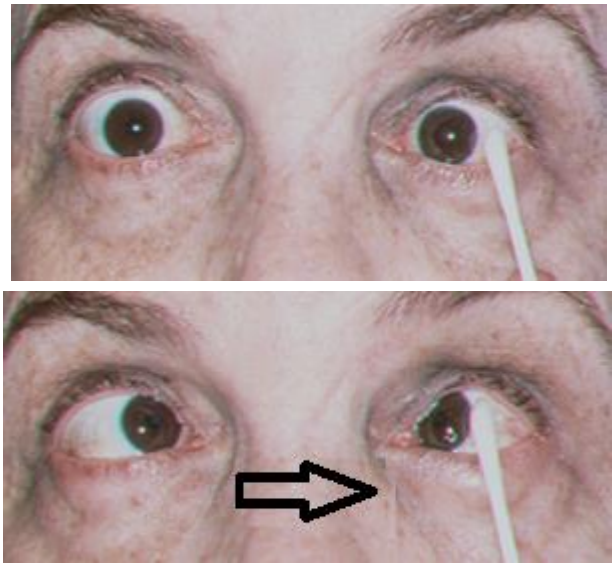
**OKN Drum:** Another way to test for a saccade is to use the OKN drum (see photo right). Slowly rotate the drum to stimulate a smooth pursuit eye movement that follows the image on the drum. After the image passes there will be a redress saccade in the opposite direction to pick up fixation and the next image. Observe for the saccadic redress movement that is opposite to the rotation of the drum. OKN testing is useful in uncooperative patients and young children. *Hey that's a photo of me 20 or so years ago.*



*Remember: Ability to produce a fast eye movement indicates the agonist muscle function is intact while lack of a saccade indicates rectus muscle paresis.*

## Force generation test

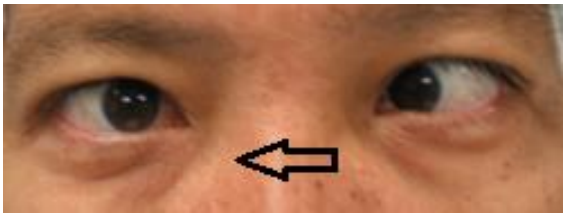
**Left 6th Nerve Palsy**



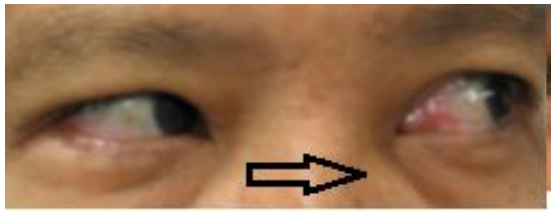
This tests muscle function as it allows the examiner to actually feel the strength of the rectus muscle. Topical anesthetic is applied and the eye fixated by forceps or a cotton applicator placed at the limbus. Note upper right photo shows a cotton tip applicator at the limbus left eye. The patient is then asked to look against the cotton tip to test the agonist muscle. In the lower right photo the patient is asked to look left (abduct) to test the lateral rectus muscle function. The examiner pushes the eye in against the lateral rectus pull. There is a 6th nerve palsy so the lateral rectus muscle is weak and cannot abduct the eye even to midline. The examiner feels no pull from the lateral rectus muscle which indicates a very weak left lateral rectus muscle.

***Lid fissure widening a subtle sign of muscle palsy:*** When a patient with a rectus muscle palsy tries to look in the field of action of the weak muscle, per Sherrington's law the antagonist is inhibited and relaxes. Because the agonist cannot contract and the antagonist is relaxed orbital pressure pushes the eye out slightly causing slight lid fissure widening. The side-by-side photos below are of a patient with a right 6th nerve palsy. Note the right lid fissure widens slightly on attempted abduction (patient looking right) as compared to adduction (patient looking left). Ok – agreed the narrowing right lid fissure is very subtle, but this is the complete opposite pattern from restriction where there is lid fissure narrowing when the eye attempts to move into the field of limited duction. Catching these subtle but real differences in lid position will make you a star on boards, and the "Mack Daddy or Mack Mama" of the strabismus clinic!

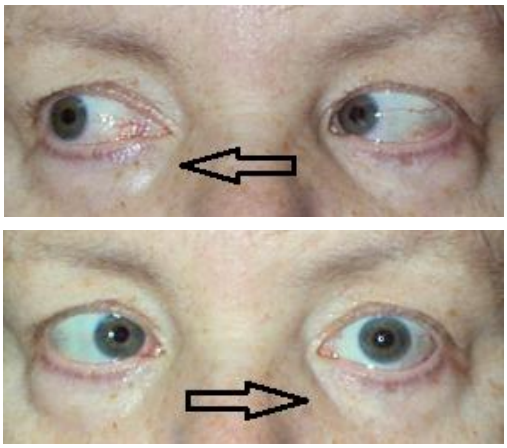
**Right 6th nerve palsy**



*Right lid fissure wide in right gaze*



*Right lid fissure narows in left gaze*



**Question:** The patient in the photos right can be best described as showing:

1. Restriction of the left eye to abduction
2. Paresis of the left lateral rectus muscle
3. Tight left medial rectus muscle
4. Limitation of abduction left eye

**Answer:** We don't know if there is restriction or paresis causing limited abduction as there is no information regarding forced ductions or rectus muscle function. Also there are no significant lid fissure changes. So, it is best just to describe the motility as limitation of abduction left eye so # 4 is correct.

**Question:** Patient above shows limited abduction left eye. How would you determine the lateral rectus muscle function?

1. Forced duction testing
2. Ability for the left eye to abduct past midline
3. Ability to generate an abduction saccade
4. Active force generation test
5. Both 3 and 4

**Answer:** Forced ductions test for restriction and does not test muscle function so # 1 is wrong. Ability to move past midline indicates degree of limitation but gives not distinguish between restriction and muscle paresis so # 2 is wrong. Both ability to generate a saccade and active force generation directly tests muscle function so # 5 is correct.

**Question:** Patient above with limited abduction shows a good abduction saccade before the eye stops abruptly. Patient cannot cooperate with forced duction testing. What is the cause of the limited abduction?

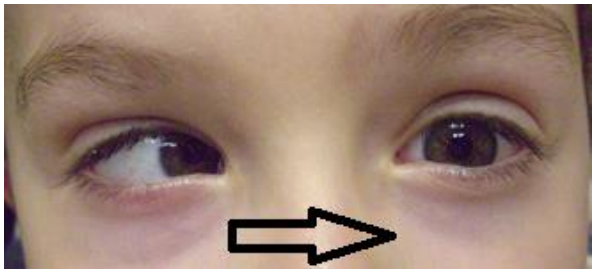
1. Lateral rectus paresis
2. Restriction from a tight medial rectus
3. Paresis of the lateral rectus muscle and tight medial rectus muscle causing restriction
4. Cannot determine without forced duction testing

**Answer:** The presence of an abduction saccade indicates good lateral rectus muscle function- no paresis so # 1 and # 3 are wrong. The pattern of fast eye movement that stops abruptly is the dog on a leash sign and indicates good rectus muscle function with restriction so # 2 is correct. Note: Forced duction testing is not necessary to diagnose restriction when you see the dog on a leash sign, however, forced ductions are helpful to verify the diagnosis.

**Note**: Often the antagonist of a paretic muscle will develop secondary contracture and become tight over several months thus adding restriction in addition to paresis as the cause for limited ductions.

## Ocular Compensatory Face Turn

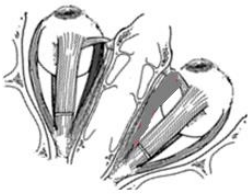
Patients with fusion potential and strabismus caused by restriction or paresis will move their head to find a position where the eyes are in alignment to obtain binocular fusion, i.e., a compensatory face turn. This is actually a compensatory gaze preference to a positon where the eyes are aligned. The photos below show a patient with a left lateral rectus palsy. There is a right gaze preference, and consequent left face turn to align the eyes and obtain binocular fusion.



**Treatment of Face Turn:** To correct the face turn move the eye with limited ductions into primary position. If there is restriction, release the cause of the restriction to improve ductions and to allow the eye to come to primary position. If the limited abduction is caused by a rectus palsy then transposition of viable muscle to the agonist would be needed to bring the eye to primary position (see below P 139). In cases of rectus muscle palsy with secondary contracture of the antagonist both transposition of viable muscle to the agonist and recession of the antagonist is indicated.

**Note:** Nystagmus with an eccentric null point is another ocular cause for a gaze preference and compensatory face turn. See chapter 11 page 154 for treatment of face turn caused by nystagmus.

**Question:** Drawing depicts the patient fusing with the eyes in left gaze and a face turn right looking at the red star. There is limited abduction right eye that causes an esotropia and diplopia that increases in right gaze. The abduction saccade is intact OU. What is causing the strabismus?



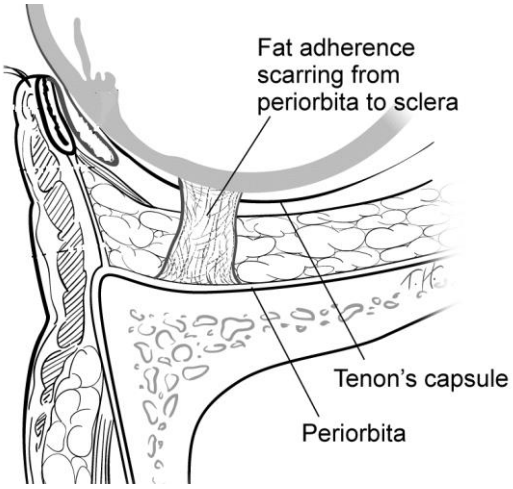
1. Right lateral rectus palsy
2. Tight right medial rectus muscle
3. Cannot determine need forced ductions

**Answer:** Limited abduction with good abduction saccade indicates muscle strength is good (not a palsy) so #1 is wrong- no palsy. Because intact saccades associated with limited ductions indicates a restriction not a palsy the limited abduction in right eye must be from a tight right MR muscle so the answer is # 2. Do not need forced ductions. This patient has a compensatory face turn to the right keeping the eyes to the left in order to avoid diplopia and fuse. Correct the esotropia in right gaze and the face turn by recessing the right MR. This will improve abduction and bring the right eye to primary position.

## Management of Specific Restrictive Disorders

For patients with diplopia from restriction, prism glasses are usually not very effective because of the incomitant pattern. Only in cases of very mild restriction does the use of prism glasses have a real chance of making the patient happy. In most cases surgery is necessary to release the restriction.

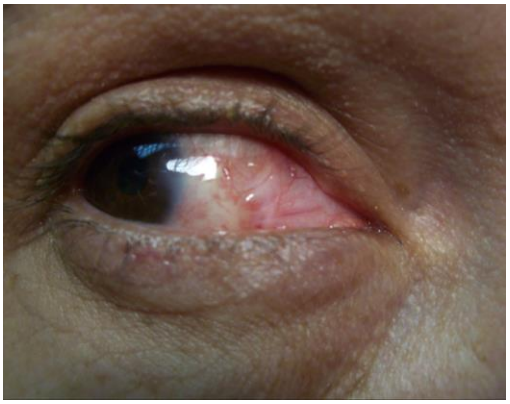
### Fat Adherence and Periocular Adhesions




Fat adherence to the extraocular muscles and/or globe is an important cause of restrictive strabismus. Drawing right shows an inferior adhesion from periorbita to the globe that restricts elevation. The best treatment for fat adherence and periocular scarring is prevention. Periocular surgery such as oculoplastic, strabismus, and retinal buckle surgery should be performed carefully to preserve posterior Tenon's capsule that separates orbital fat from the globe. When isolating an extraocular muscle keep the dissection close to the muscle – do not violate posterior Tenon's capsule as this exposes orbital fat causing adhesions to the globe.

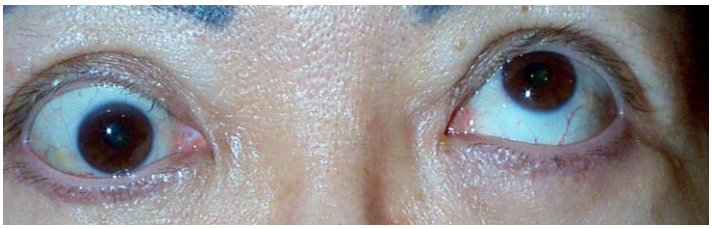
Surgical correction of fat adherence is difficult as recurrence of scar is common. Perform progressive dissection combined with forced ductions until the restriction is released. After adhesions are removed try to close the tear in the Tenon's capsule with 7-0 vicryl suture. Use of mitomycin C is not helpful to prevent recurrence and can even lead to further scarring.

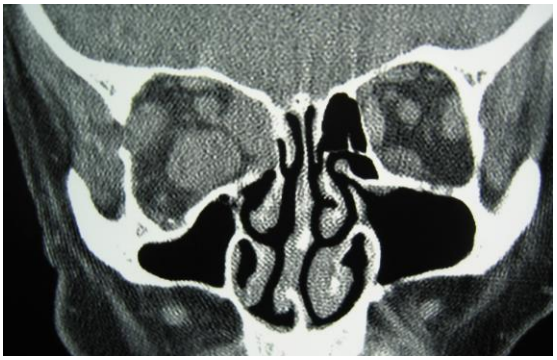
**Tight scarred conjunctiva –** Scarring and shortening of the conjunctiva can cause restriction. Conjunctival scarring after pterygium surgery can cause restrictive strabismus. Photo right shows nasal scarring after multiple pterygium surgeries and use of mitomycin C. This patient had an esotropia caused by tight nasal conjunctiva.



Treatment is complete removal of the scar and replacement of the missing tissue with a autologous conjunctival graft or an amniotic membrane transplant. Recession of the corresponding rectus muscle is usually required such as a MR muscle recession for esotropia associated with nasal conjunctival scarring after pterygium surgery.

## Thyroid autoimmune strabismus





Thyroid eye disease is an autoimmune disease that affects the thyroid gland and the extraocular muscles and periorbital tissue. The first phase is inflammatory and usually lasts a year to two years. After the inflammatory phase there is a fibrotic phase as the extraocular muscles scar and contract. Strabismus usually occurs during the fibrotic phase as the extraocular muscles become fibrotic and stiff. Various extraocular muscles can all be involved and the disease can be fairly symmetrical or very asymmetric. The inferior and medial rectus muscles are most frequently involved. Therefore, hypotropia with restricted elevation and esotropia with restricted abduction are the most common strabismus. Photo upper right shows tight right IR and restriction of upgaze. Photo of the coronal orbital CT scan shows the corresponding enlarged medial and inferior recti in the right orbit. Surgery is performed once the acute inflammatory phase has subsided and the angle of deviation is stable, usually after a minimum of 6 months. Recessions should be performed on the tight muscles, and resections are avoided when possible. Non-absorbable sutures are recommended to avoid the common scenario postoperatively of late overcorrection due to stretched insertion scar or dehiscence.

**Congenital fibrosis syndrome** is associated with tight fibrotic extraocular muscles with the medial and inferior rectus muscles more often involved. Photo right shows a child with congenital fibrosis with tight right IR and left MR muscles. It is usually inherited as an autosomal dominant trait. It is one of the congenital cranial dysinnervation disorders (CCDDs), with the oculomotor, trochlear and abducens nuclei found to be maldeveloped. Treatment is large bilateral MR recessions (usually 7to 8 mm).

**Brown's syndrome –** This is a form of restrictive strabismus caused by an inelastic superior oblique muscle. The tight SO tendon causes restriction of elevation in adduction. Please see chapter 11 page 147 for more details.

## Treatment of Rectus Muscle Paresis

The surgical treatment depends on the severity of the paresis. For mild paresis with muscle strength at 50% to 70% then standard recession and resection or plication surgery will work. If, however, the paresis is profound, standard surgery will not hold alignment and transposition of viable muscle to the paretic muscle is required. It is important to assess the amount of rectus muscle function prior to surgery.

### Sixth Nerve Paresis

Cranial nerve six innervates the lateral rectus muscle. A sixth nerve paresis will produce an esotropia greater for distance because of lack of divergence. The esotropia is incomitant increasing to the side of the paresis as abduction is reduced and because of Hering's law the yoke muscle the medial rectus of the good eye will appear to over act. An acquired sixth nerve palsy from a vascular or traumatic cause should be observed for 6 months to allow for lateral rectus muscle function to recover before attempting surgery. Use monocular occlusion or try press-on prisms to treat diplopia while waiting for recovery of muscle function. **Use of Botox:** Some have advocated injection of the ipsilateral medial rectus muscle with botulinum toxin to prevent secondary muscle contracture. Botox has not shown to improve muscle function recovery or reduce need for surgery. Also there are significant possible complications of Botox injection including ptosis, induced vertical strabismus and globe perforation. Because of this I do not advocate Botox injection for the acute management of sixth nerve palsy.

## Sixth Nerve Paresis – Good LR Function

Patient in the photos below has a partially recovered vascular right sixth nerve paresis with virtually full ductions and good lateral rectus function. He has a small esotropia worse for distance and increases in right gaze. Note compensatory face turn to the right with a left gaze preference to obtain binocular fusion and avoid diplopia. Because the esotropia is small in primary and ductions are full a good strategy is to weaken (recess) the left MR muscle. By weakening the left MR muscle you match the slight weakness of the right LR muscle (yoke muscles) improving the incomitance. Weakening the yoke muscle of the paretic muscle will improve the incomitance if the paresis is mild. One could also slightly tighten (Wright central plication) the left lateral rectus which would also limit adduction left eye thus correcting the incomitance.


**Question:** A patient presents with an acute traumatic bilateral 6th nerve paresis. What would you do?

1. Recession medial rectus OU
2. Recession one medial and resection one lateral
3. Faden ipsilateral medial rectus
4. Wait at least 6 months before considering surgery.

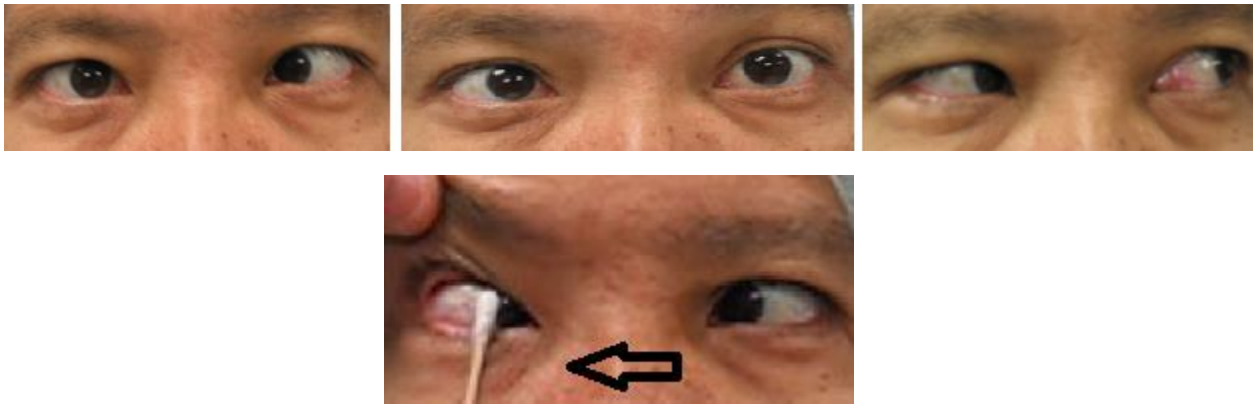
**Answer:** Must wait 6 months for muscle function improvement so answer is # 4.

**Question:** A teenager sustained a traumatic left 6$^{th}$ nerve palsy 6 months ago. There was some recovery, but now the patient presents with diplopia and an ET 8 in primary which increases to ET 16 in left gaze. There is a good abduction saccade OU, but -1 abduction OS. Which is FALSE?

1. Uncrossed diplopia in left gaze
2. Residual mild left 6$^{th}$ paresis
3. Lateral rectus function is very poor so patient needs a transposition surgery
4. Recession of right medial rectus muscle would help the incomitance.

**Answer:** Esotropia gives uncrossed diplopia and since the esotropia increases in left gaze #1 is true. There pattern of strabismus indicates good LR muscle function and mild residual left 6th nerve paresis so #2 is true. Lateral function is good because there is a normal abduction saccade, so #3 is false and is the answer. Because of the good LR muscle function a recession of the yoke to the paretic muscle the right MR muscle would reduce the ET that is greater in left gaze and improve the incomitance and #4 is true.

## Six Nerve Palsy – Poor Lateral Rectus Function



Patient in the composite photos above has a complete right sixth nerve palsy and virtually no lateral rectus muscle function. There is no abduction saccade and see photo above showing the force generation test showing no lateral rectus function. Surgical treatment of a complete paralysis is a transposition surgery moving all or part of vertical recti to the lateral rectus muscle (see below). The medial rectus muscle is usually tight from secondary contracture so an ipsilateral medial rectus recession is usually indicated.

## *Summary of the Management of 6th NP*

### GOOD LR FUNCTION

| | |
|---|---|
| Unilateral ET < 20 | Recess contralateral MR<br>Wright central plication contralateral LR |
| Bilateral ET > 20 | Bilateral MR recessions |

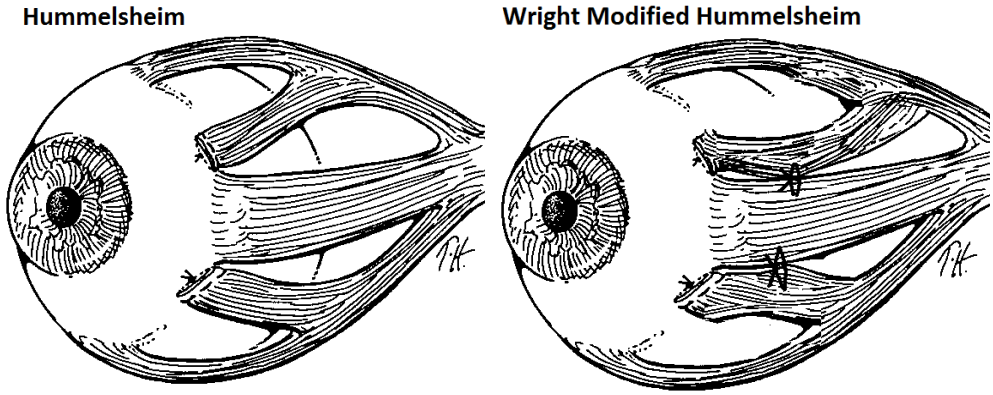
### POOR LR FUNCTION

Vertical muscle Transposition to LR - Partial tendon (Hummelsheim or Jensen or full tendon transposition with ipsilateral MR recession. Full tendon transfer of SR and IR to the LR can be done but anterior segment ischemia is a risk. (See next page)

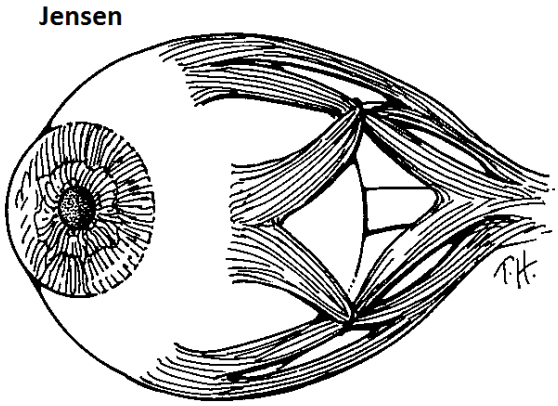
## Partial Rectus Muscle Transposition Procedures

Partial muscle transpositions Hummelsheim and Jensen spare the nasal anterior ciliary vessels of the superior and inferior rectus muscles. This is a significant advantage over the full tendon transposition. Anterior segment ischemia is a possible complication of transposition surgery especially if the ipsilateral medial rectus muscle is recessed. I prefer a partial tendon transposition to preserve anterior ciliary arteries and reduce the risk of anterior segment ischemia.

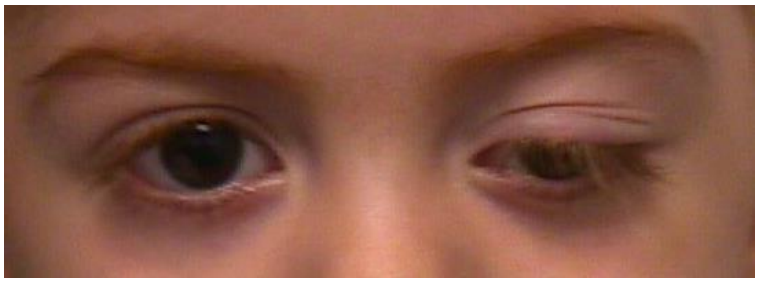
**Hummelsheim –** Temporal 1/3 to 1/2 of the superior and inferior recti are disinserted and transposed to the lateral rectus insertion as shown in the drawing below. I modify the Hummelsheim by suturing the transposed vertical muscle to the lateral rectus muscle 5 mm posterior to the insertion.




**Jensen Union -** Transposition unites the temporal aspect of the superior and inferior recti to the lateral rectus using a non-absorbable suture without needing to disinsert the muscle.



### Third nerve palsy



Etiology of third nerve palsy can be congenital or acquired (e.g., trauma, aneurysm, or tumor). The palsy may involve all of the extraocular muscles except the superior oblique and lateral rectus muscles.

A complete third nerve palsy typically presents with the eye down and out with ptosis as seen in the photo above right of a patient with a congenital left third nerve palsy. With a complete third nerve palsy the eye is virtually fixed in abduction as there is no elevation, depression, or adduction and only abduction is intact. On attempted downgaze you can see slight intorsion indicating that the superior oblique muscle is functioning.

**Treatment:** Treat a partial third nerve paresis with the good medial rectus muscle function by an ipsilateral lateral rectus muscle recession and a medial rectus muscle resection or plication. Add an ipsilateral superior oblique weakening procedure if a hypotropia is present. If there is a complete third nerve palsy and muscle function is poor, the recess-resect procedure does not work well in the long term. In these cases split the lateral rectus in two. Then transpose the superior half nasal to the superior rectus insertion, and transpose the inferior half of the lateral rectus nasal to the inferior rectus insertion. Add a superior oblique tenotomy if there SO function and hypotropia. Be careful about correcting associated ptosis in a patient with a complete third palsy. Because these patients have poor superior rectus function a frontalis sling procedure runs the risk of creating corneal exposure. Best to undercorrect the ptosis repair if there is no Bell's phenomenon.

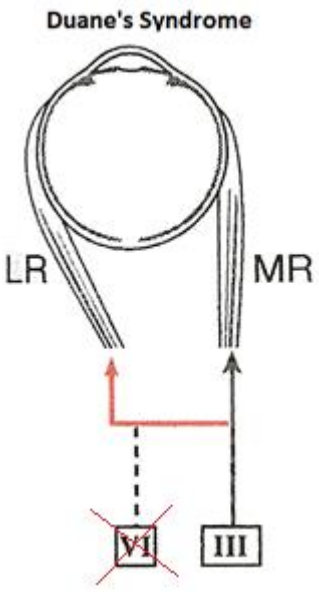
**Question:** Patient has traumatic left 6$^{th}$ NP for 6 months and now has –3 abduction OS and cannot generate an abduction saccade OS. One acceptable surgical approach is:

1. Left Hummelsheim transposition and MR recession
2. Right MR recession
3. Left MR recession
4. Left MR recession and LR Resection

**Answer:** There is a non-recovering left 6$^{th}$ NP with poor LR function as there is limited abduction and no abduction saccade OS. Because the left LR function is poor a left transposition is necessary so the answer is #1.

# Chapter 11 Strabismus Syndromes

## Duane's Syndrome




Duane's syndrome is a congenital agenesis of the sixth nerve nucleus with the medial rectus branch of the third nerve splitting. Part of the medial rectus nerve goes appropriately to the medial rectus but part aberrantly innervates the lateral rectus muscle (see drawing right). Drawing right shows third nerve going to the medial rectus (black arrow), but also splits to innervate the lateral rectus (red arrow). Note the sixth nerve and nucleus are absent. Because the LR and MR are innervated by the same MR nerve (inferior division of CN 3) they both contract on adduction. Because there is no sixth nerve there is an abduction deficit.

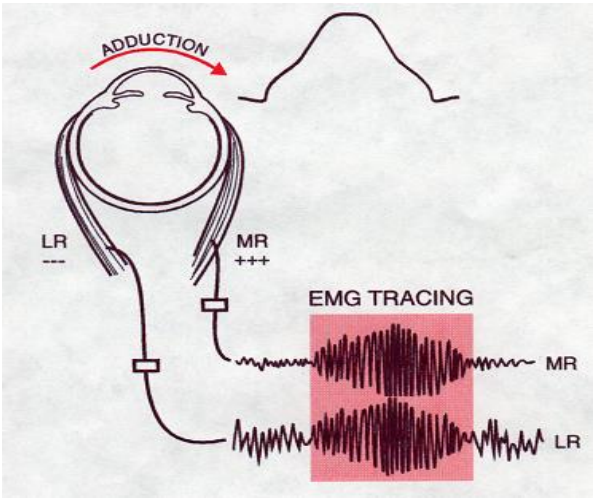
### Key Clinical Characteristics

- Abduction deficit
- Lid fissure narrowing on adduction

### Abduction Deficit

Because there is no cranial nerve 6 abduction is limited in virtually all Duane's syndrome patients. However, different than a true sixth nerve palsy in Duane's syndrome the lateral rectus is innervated by part of cranial nerve 3 so the lateral is not "dead" and has tone. Rarely a patient with Duane's syndrome type II will have some abduction because of lateral rectus tone.

### Lid Fissure Narrowing on Adduction




Since the medial and lateral recti muscles are innervated with the same nerve, both muscles contract on adduction. The electromyograph (EMG) shown in the diagram right shows co-contraction with both medial and lateral recti receiving innervation on adduction. This co-contraction of the medial and lateral rectus muscles pulls the eye posteriorly causing lid fissure narrowing on adduction, a key clinical finding of Duane's syndrome. Thus the alternative name "Duane's Retraction Syndrome."

## Classification

Three forms of Duane's syndrome have been described with type I most common. Type I has minimal third nerve innervation to the LR muscle, and type III has equal innervation of MR and LR by the third nerve. Type II is very rare and may not even exist except in the old literature. Type II supposedly has intact sixth nerve innervation and intact abduction (not really Duane's). Duane's syndrome is usually unilateral but can be bilateral.

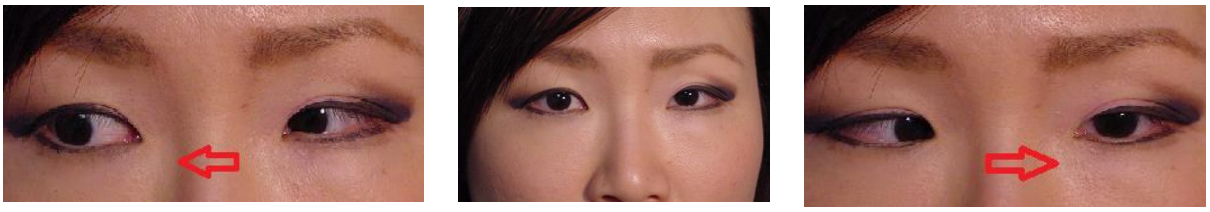
- Type I: limited abduction with intact adduction
- Type II: limited adduction with intact abduction (rare – exists?)
- Type III: limited abduction and limited adduction

*Synergist divergence* is a rare striking motility pattern caused by the lateral rectus getting most of the innervation from the MR nerve. When the "good" eye abducts the Duane's eye also abducts.

*Because patients with Duane's syndrome have binocular fusion, albeit with a face turn they do not get amblyopia unless there is another cause such as hyperopic anisometropia. Patients with Duane's syndrome usually do not complain of diplopia as they suppress when tropic. Thus they can fuse and suppress, as do virtually all patients with childhood onset intermittent strabismus (e.g. intermittent XT, Brown's syndrome, and congenital SOP).*

### Duane's Syndrome Type I

Composite photos below show a left Duane's syndrome with -4 limitation of abduction and there is mild lid fissure narrowing left eye on adduction. The full face photo center shows a slight right gaze preference with a face turn left. With the eyes in right this allows the Duane's eye (left eye) to remain in its position of rest in adduction. The normal right eye moves freely to line up with the Duane's eye stuck in adduction. Because Duane's syndrome is congenital these patients learn to suppress the diplopia when tropic, yet they have high grade stereo acuity when aligned with a face turn.



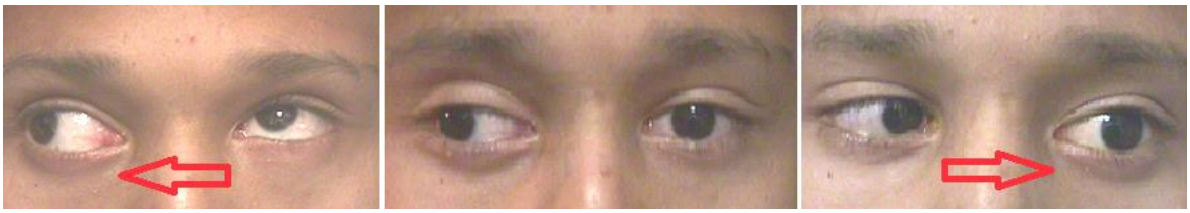
**Question:** The patient above shows left eye abduction deficit with lid fissure narrowing left eye on adduction. All of the following are correct EXEPT:

1. Violation of Sherrington's law of agonist / antagonist
2. Abduction deficit left eye from agenesis of left abducens nucleus
3. Lid fissure narrowing on adduction due to aberrant innervation of lateral by medial rectus nerve
4. Amblyopia LE

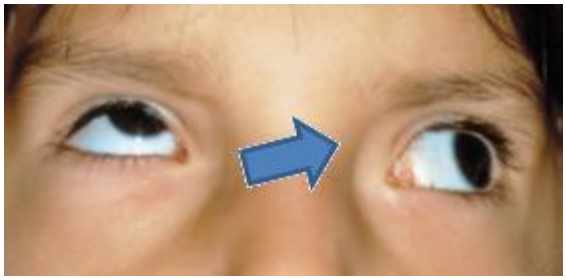
**Answer:** All are correct except # 4. Patients with Duane's syndrome have high-grade stereopsis with a compensatory face turn so they typically do not have amblyopia unless there is anisometropia.

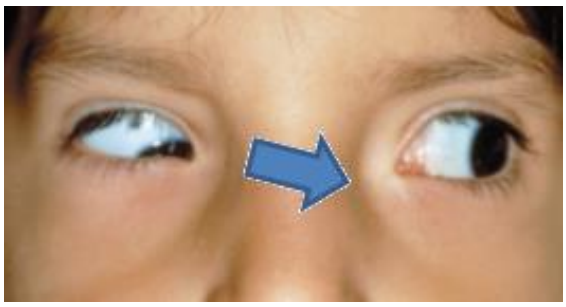
## Duane's Syndrome Type III

Photos below show Duane's III left eye. Note left eye does not adduct but up-shoots in attempted adduction with lid fissure narrowing. Abduction is better than adduction but still limited. All Duane's syndromes are misdirection of third and six cranial nerves of various patterns of innervation. The patient below probably represents: part of the MR nerve goes to SR muscle and part goes to the LR muscle causing elevation of the left eye on attempted adduction and co-contraction of MR and LR causing limited adduction and globe retraction with lid fissure narrowing. Note- fixing with paretic left eye in primary position.



**Upshoots and downshoots:** "Upshoots" and "downshoots" on attempted adduction are often seen in patients with Duane's syndrome especially type III. Photos right show right Duane's syndrome with limited adduction, and upshoot (upper photo) and downshoot (lower photo). In most cases it is due to co-contraction of the lateral rectus muscle which leashes the eye up and down. Recessing the lateral rectus muscle reduces the vertical shoots. A "Y" split of the LR muscle and recession has been reported by your author as also being an effective treatment. Occasionally the vertical shoot is caused by an innervational misdirection with the MR nerve going to a vertical muscle as seen in composite of Duane's type III above.





## Treatment of Duane's Syndrome

The indication for surgery is a face turn. As is true with other types of strabismus caused by an eye stuck in an eccentric position the surgical goal is to move the stuck eye into primary position. For Duane's type I with esotropia in primary position recess the ipsilateral MR muscle. Because the LR has tone from aberrant third nerve innervation a simple recession of the MR will result in a stable correction. A transposition of the vertical rectus to the LR muscle has been suggested by some authorities but is not necessary as it is for a true 6th nerve palsy. For a type III exotropia Duane's syndrome recess the LR muscle. In general do not resect or tighten muscles because of co-contraction.

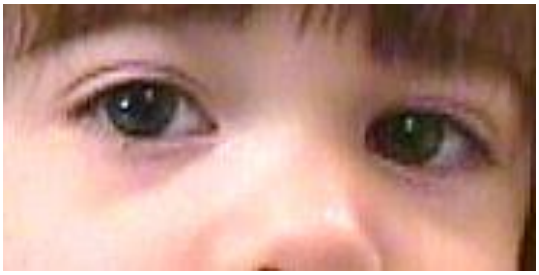
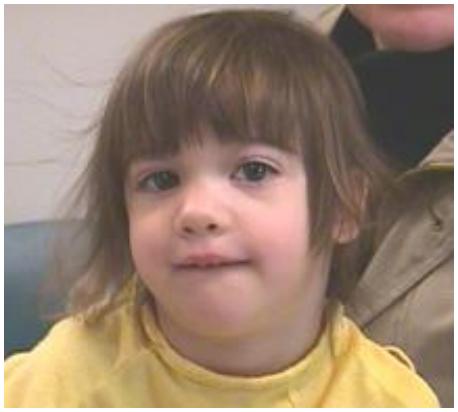
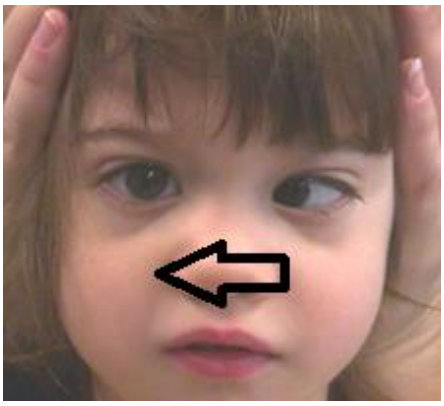


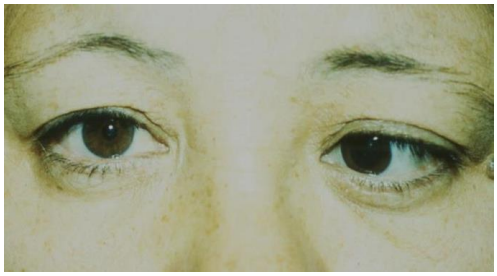

Photo above left shows left Duane's type I with left eye stuck in adduction and right gaze preference for fusion (compensitory face turn left). Right shows postoperative alignment after left MR recession 5.5 mm. Note by recessing left MR muscle this moves the left eye into primary position and the right eye with full movement follows so both eyes are aligned in primary position correcting the face turn.

**Question:** For the patient below which is correct?

1. Right type 1 esotropia Duane's syndrome
2. Gaze preference left with face turn right allows high-grade stereo acuity
3. Correct the face turn by recessing right MR muscle
4. Photo right shows esotropia in right gaze and in this gaze the patient suppresses diplopia
5. All of the above

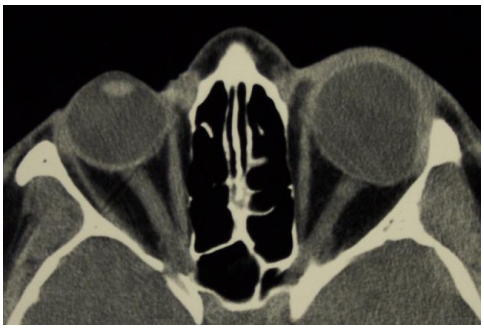



**Answer:** All the above are correct # 5. Note that you can correct the face turn by moving the Duane's eye (right eye) to the right into primary position. A simple right MR recession (about 5 to 6 mm) will work well. The muscle tone of the LR muscle provided by aberrant third nerve innervation will hold the eye in position and transposition of vertical recti to the LR muscle is not needed.

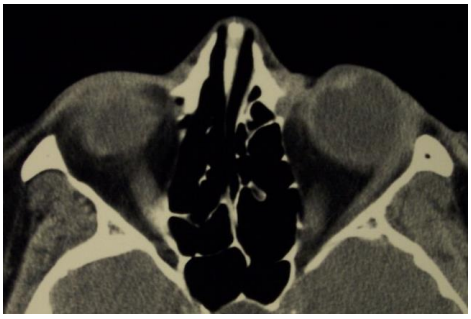


## High Myopia Esotropia and Hypotropia ("Heavy Eye Syndrome," "Sagging Eye")

Axial high myopia can cause acquired esotropia and hypotropia with symptoms of diplopia (photo upper right). This occurs because the macular staphyloma expands superotemporally pushing the LR muscle down and the SR muscle nasally. With the LR displaced down it becomes more of a depressor and less of an abductor and the SR displaced nasally becomes even more of an adductor and less of an elevator This displacement of the lateral and superior recti result in limited elevation and abduction.



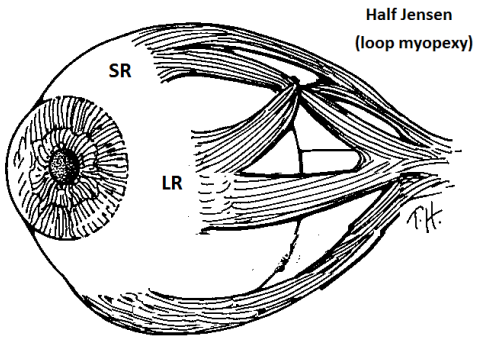
CT upper right shows cut through middle of eye and left staphyloma showing the horizontal recti except there is no LR muscle as it is displaced inferiorly.



CT lower right is a cut below showing the infra-placed LR muscle

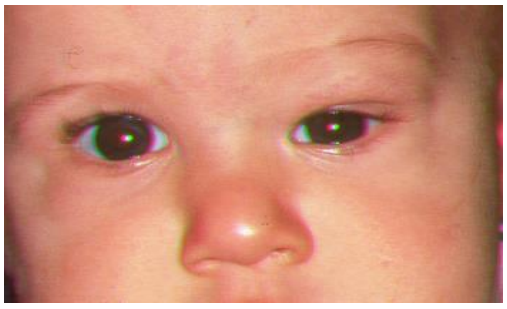
### Key Clinical Points

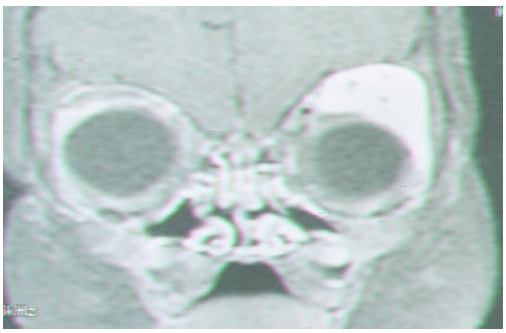
- Axial myopia (>15 D)
- Esotropia and hypotropia
- Limited elevation and abduction
- LR displaced down and SR displaced nasally

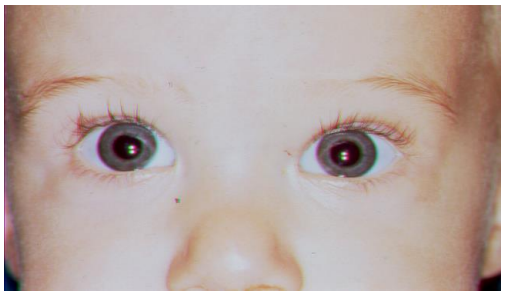



**Treatment:** The surgical treatment described by Yokoyama is based on moving the displaced LR and SR muscles back to where they should be: SR temporal, LR up. This can be done with a half Jensen (loop myopexy) procedure as shown drawing right. The SR and LR muscles are split in two then the temporal half of SR is looped to the upper half of the LR with a non-absorbable suture (5-0 Mersilene). An ipsilateral MR recession is usually also required.

## Elevation Deficit







**Question:** Infant in photo right has a left hypotropia with limited elevation. What do you want to do?

1. Recess left IR muscle
2. Forced ductions at the time of surgery
3. Observe
4. MRI of orbit

**Answer:** Note that there is a left hypotropia but there is also a left ptosis and an upper lid bulge. The answer is obtain MRI of orbit. Image right show a superior temporal orbital hemangioma. The patient was given an intralesional corticosteroid injection.

Photo right shows upper lid mass gone and resolution of strabismus after steroid injection of the hemangioma.

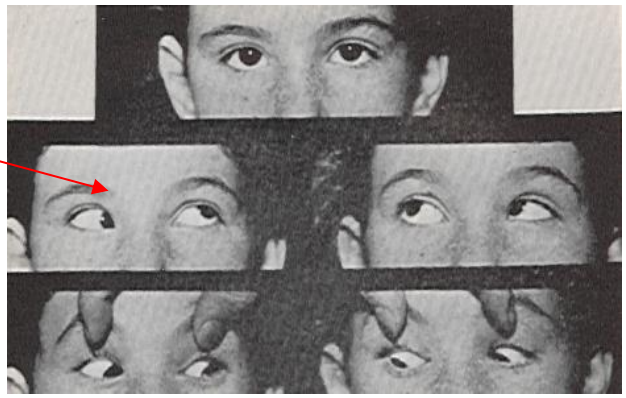
Remember: Strabismus can be from orbital or neurological disorders. If not a typical garden-variety strabismus then obtain further investigation and get the MRI scan.

### Causes of Limited Elevation

| Congenital | Acquired |
|---|---|
| Brown's Syndrome | Floor Fracture |
| Inferior Oblique Palsy | Local Anesthesia Myotoxicity |
| Double Elevator Palsy (MED) | Orbital Mass |
| Cong. Third Nerve Palsy | Thyroid Autoimmune Disease |

## Brown's Syndrome

In 1950 Harold Brown first published a series of patients with restriction of elevation in adduction. This Brown's Syndrome is simply a restriction of elevation in adduction. This restriction is in most cases due to an inelastic superior oblique muscle tendon complex. The tight superior oblique restricts elevation that is worse in adduction. It can be congenital or acquired, usually unilateral but it can be bilateral. Note composite photos from Brown's original article showing right congenital Brown's syndrome with severe limitation of elevation in adduction.



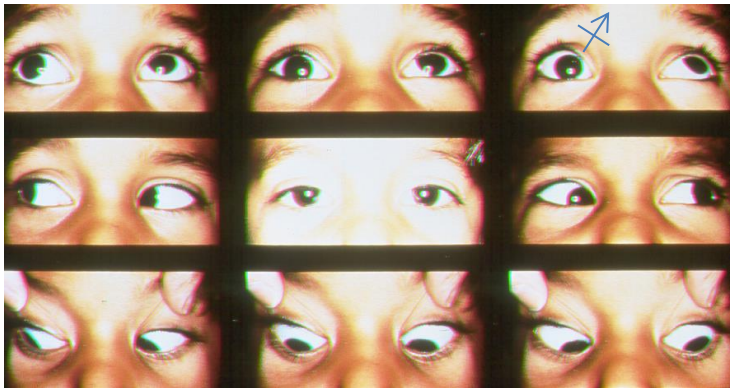
## Clinical Characteristics

- Small hypotropia (usually < 5 PD)
- Limited elevation in adduction
- Little if any SO overaction
- Near normal elevation in abduction
- Often abduction in upgaze ("Y" pattern)
- Restricted forced ductions to elevation in adduction
- Compensatory chin elevation

## Congenital Brown's Syndrome

Even though congenital Brown's syndrome is present at birth it is often first noticed in later in childhood. With a chin elevation patients can achieve binocular fusion and high-grade stereo acuity. In most cases the etiology is a tight superior oblique muscle tendon complex. Your author published that the best explanation for the pattern of strabismus is an inelastic SO muscle tendon complex. Perhaps it is a fibrosis of the superior oblique muscle. Rarely an anomalous inferior congenital band or anomalous location of a muscle can cause the pattern of Brown's syndrome.

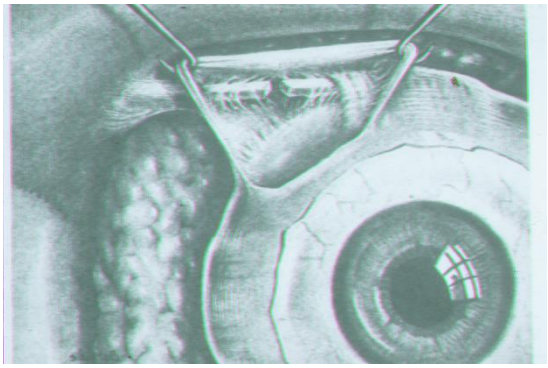
Note the composite photos right show a right Brown's syndrome with limited elevation worse in adduction than abduction (arrow). Note good alignment in primary position, no SO overaction, and an XT in up-gaze.



## Treatment of Congenital Brown's Syndrome

If the Brown's syndrome is mild and the patient is fusing well with a small compensatory chin elevation there is no rush to do surgery. In fact I only operate if the Brown's syndrome is severe and there is a significant chin elevation and I invented the surgery. Congenital Brown's syndrome is caused by a tight superior oblique muscle tendon complex, so surgery is based on relaxing the tight SO tendon.

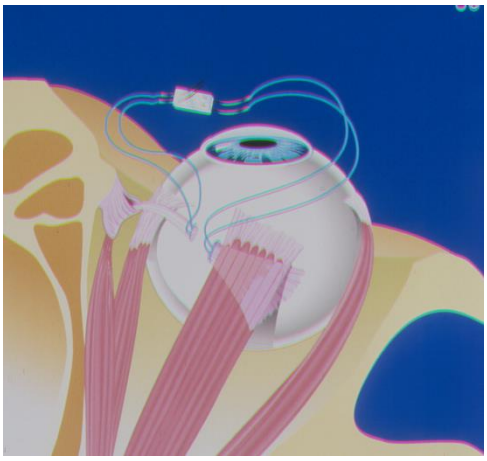
SO tenotomy (drawing right) had been used in the past but it resulted in uncontrolled separation of the cut tendon ends with 80% of cases resulting in a consecutive SO palsy. Most patients were worse off after surgery and for many years most authorities warned against operating on Brown's syndrome. This procedure has been abandoned for the treatment of Brown's syndrome.

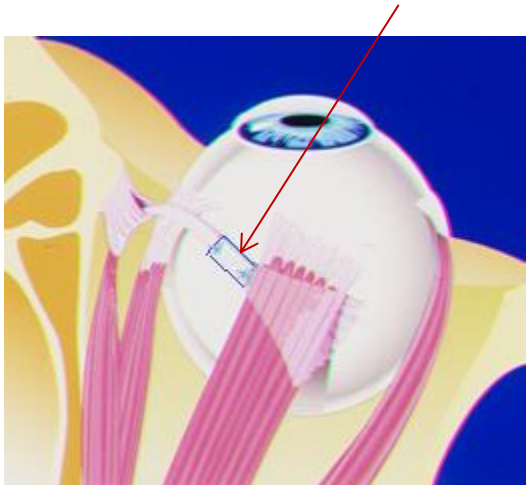


Berk Superior Oblique Tenotomy

In 1992 your humble author published a new procedure that elongates the tendon called the "SO silicone tendon expander." A 5 mm segment of a retinal 40 band is placed between the cut ends of the SO tendon with a success rate of over 80%. The Wright SO silicone expander has become a popular choice for Brown's syndrome. Drawing below left shows sutures attached to cut ends of SO tendon and needles being passed through a segment of a silicone retinal band. Drawing right shows the segment of silicone band in place elongating the SO tendon (arrow).

**Wright KW**, Min BM, Park C. Comparison of superior oblique tendon expander to superior oblique tenotomy for the management of superior oblique overaction and Brown syndrome. J Pediatr Ophthalm Strabismus 1992;29(2):92-97.





**Question:** What is true about congenital Brown's syndrome?

1. Early surgery is critical.
2. Amblyopia is common.
3. Sensory adaptation like Duanes's syndrome and intermittent exotropia – high-grade stereo acuity and cortical suppression
4. Large hypotropia in primary position

**Answer:** Patients with Brown's syndrome have excellent binocular fusion with a chin elevation so we can wait on surgery and amblyopia is rare. The answer is # 3 as with the chin elevation there is high grade stereo acuity, and with upgaze when hypotropic the patient can suppress diplopia.

## Acquired Brown's Syndrome

Inflammation in the area of the trochlea can cause restriction of SO tendon movement and cause acquired Brown's syndrome usually with the symptom of diplopia. Acquired Brown's syndrome is often intermittent and some patients can feel a bump and hear a click in the area of the trochlea. Rarely the physician can feel and hear the click with a stethoscope as the attempts to move up and in. Many acquired Brown's will resolve spontaneously so in general, surgery is not indicated. The work up should include orbital imaging.

### Causes of Acquired Brown's Syndrome

| | |
|---|---|
| **Inflammatory** | |
| Primary tenosynovitis | Rheumatoid arthritis |
| Sinusitis | Upper respiratory infection |
| Peri-trochlear inflammation | Trochleitis |
| **Orbital Trauma** | |
| Superior nasal orbit trauma | Canine Tooth syndrome |
| **Superior Nasal Mass** | |
| Orbital neoplasm | Nasal glaucoma implant with large bleb |
| **Iatrogenic** - Superior oblique tuck | Retinal band that tightens SO tendon |

**Question:** Brown's Syndrome is caused by an inelastic or tightness of the superior oblique muscle tendon complex as it passes through the trochlea- True of False?

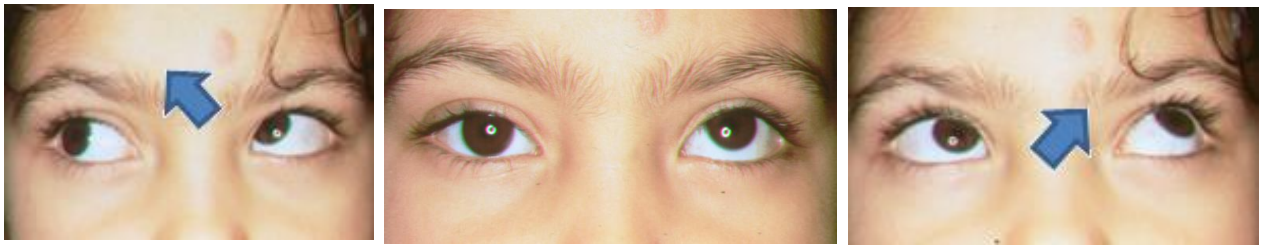
**Answer:** True. Tightness of the SO holds the back of the eye preventing elevation worse in adduction**. Forced ductions are positive** with restriction of elevation in adduction.

**Treatment of Acquired Brown's Syndrome:** Treatment is based on the cause of the acquired Brown's syndrome. For inflammatory Brown's syndrome, a trial of systemic non-steroidal anti-inflammatory agents or a local steroid injection in the area of the trochlea can be effective. Acquired Brown's syndrome of unknown etiology should be worked up with orbital imaging as a variety of local or systemic diseases around the trochlea may cause Brown's syndrome. Medical therapy, not surgery, is the treatment of choice for most cases of inflammatory Brown's syndrome.

**Canine Tooth Syndrome:** Trauma to the area of the trochlea can cause scarring of the SO tendon as it exits the trochlea. Dog bites often involve the superior nasal face thus the name "canine tooth." Canine tooth syndrome is when scarring in area of the trochlea causes a "frozen trochlea" as the tendon movement is restricted in both ways. This causes a Brown's syndrome with limitation to elevation in adduction and weakness of the SO function as muscle pull cannot be transmitted to the globe. Treatment is difficult as dissection of scar often results in more scarring.

## Double Elevator Palsy (DEP) or Monocular Elevation Deficit (MED)

Congenital elevation deficit with limitation of elevation more in adduction is most likely Brown's syndrome, however, if the elevation deficit is more in abduction like the patient in the photos below this has been historically called "double elevator palsy." This term is actually a misnomer because the cause in 80% of patients with this pattern is actually restriction from a tight IR muscle, not palsy. This was discovered by Dr. Henry Metz who performed saccadic velocity measurements and found normal upgaze saccades and a "dog on a leash" pattern of restriction. Only 20% are a true "palsy" of elevation i.e., superior rectus palsy. Monocular elevation deficit syndrome is a better term to use until the diagnosis is confirmed. Note the patient below has monocular elevation deficit right eye with the elevation deficit is worse in abduction and better elevation in adduction.

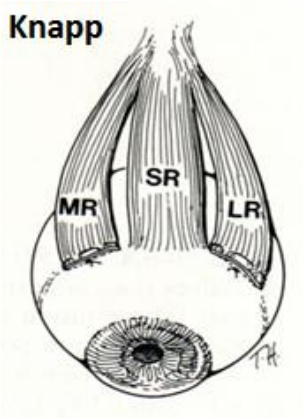


Be sure to check for an upgaze saccade. If an upgaze saccade is present this means good SR muscle function and surgical treatment is a recession of the ipsilateral IR muscle. Absence of a saccade indicates a true palsy and a muscle transposition is required. Patients with MED will have a chin elevation. With the compensatory chin elevation there is good binocular fusion with stereo acuity.

**Treatment MED:** Indication for surgery is the presence of a significant chin elevation.

**Tight IR good SR function:** Recess ipsilateral IR muscle

**True SR muscle Palsy:** The treatment of a rectus palsy is to transpose viable muscle to the paretic muscle. Full tendon transfer (Knapp procedure) or partial tendon transfer of the LR and MR up to the SR is indicated. The IR muscle is usually tight so if forced ductions are positive consider recessing the ipsilateral IR muscle. Better to perform a partial tendon transfer if recessing the IR muscle to preserve anterior segment circulation.




**Question:** 8 year old shows poor elevation OS worse in abduction since infancy and no diplopia. There is a good up gaze saccade so you should recommend a Knapp full tendon transfer: True or False?

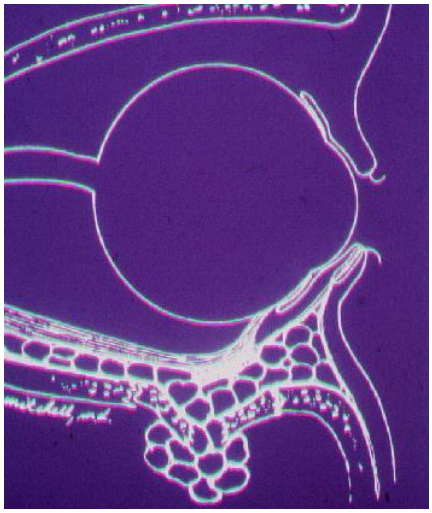
**Answer:** False. A good up gaze saccade indicated good SR function so a transposition is not indicated. This is MED secondary to a tight IR so recess the IR OS.

**Question:** Patient in photo right is 4 years old and presents with double vision and acquired limited elevation left eye. Best management is:

1) Release restriction by recessing left IR muscle
2) Knapp transposition of MR and LR to SR muscle
3) Recess right SR muscle to stop overshoot
4) MRI of orbit and brain

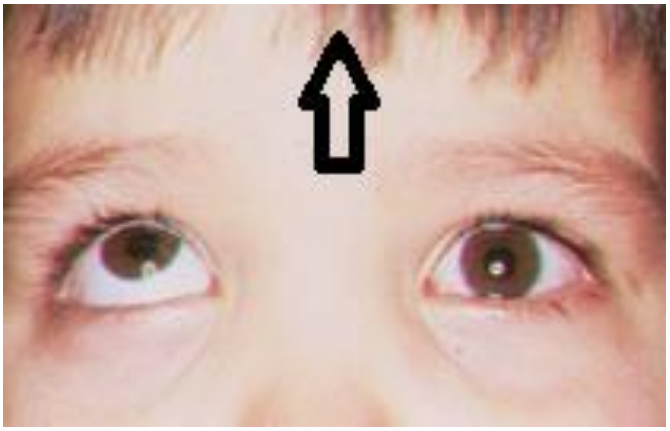
**Answer:** The symptom of diplopia is a red flag that the strabismus is acquired. Acquired strabismus with limited ductions indicates the possibility that the strabismus could be "dangerous" and there could be a brain tumor pushing on the third nerve nucleus or an orbital mass restricting elevation. The correct answer is # 4 to obtain a MRI scan and this patient actually had a midbrain glioma pushing on the third nerve nucleus. Note no information regarding presence of an upgaze saccade or forced duction therefore it is not possible to make a decision regarding surgical treatment.

## Orbital Floor Fracture - Strabismus

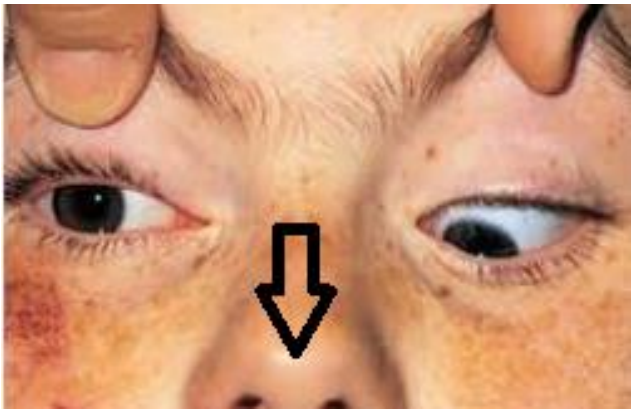


After repair of a floor fracture, vertical diplopia is common. There can be residual entrapment of the IR, scarring of inferior orbital fat or scarring to the floor implant that will interfere with globe rotation. Note that entrapment of orbital fat can cause hypotropia as the herniated fat pulls on the inferior rectus muscle (see drawing right). There are two basic types of vertical strabismus after orbital floor fracture: 1) inferior orbital restriction, and 2) functional paresis IR muscle. Motility can improve over time so best to wait several months after orbit repair to do the strabismus surgery.

**Inferior Orbital Restriction:** The most common strabismus is a hypotropia and restriction of elevation from inferior restriction either from scarring of orbital fat around the IR muscle or residual IR muscle entrapment. The inferior restriction restricts elevation. Photo right shows limitation of elevation left eye after left floor fracture. This patient had good upgaze saccade indicating good SR function and forced ductions at the time of surgery verified inferior rectus restriction. *Treatment* is do forced ductions to verify IR restriction then recess the tight IR muscle.

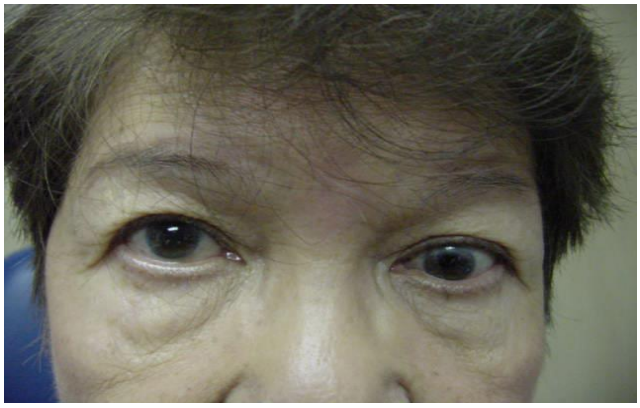


**Functional Paresis of IR Muscle:** Less common than limited elevation after orbital floor fracture is limited depression. Photo right shows a patient with a right floor fracture and limited depression right eye. These patients have apparent underacting inferior rectus, but usually intact down gaze saccades. Treatment is plication of the ipsilateral IR muscle and if the hypertropia is > 10 PD then add a recession of the contralateral IR muscle



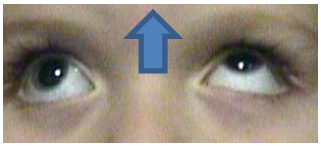
## Strabismus Caused by Anesthetic Injection

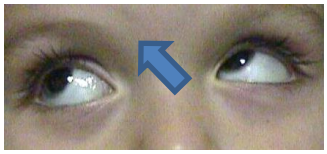
Retrobulbar injection of local anesthesia such as bupivacaine injected into an extra- ocular muscle will initially cause paralysis, and after 2 to 3 weeks the muscle will hypertrophy and overact. Anesthetic injection into an extra-ocular muscle most frequently occurs with Retrobulbar or perbulbar anesthestetic injections. The inferior rectus is most prone to inadvertent injection but any muscle can be involved. Muscle hypertrophy occurs because after local anesthetic breaks up muscle fiber integrity. When the muscle reorganizes it hypertrophies and becomes stronger. This is similar to what happens when weight lifting: you first break down muscle, then muscle hypertrophies as it reorganizes. Dr. Allen Scott from San Francisco has used injection of anesthetic into rectus muscle to strengthen the muscle to treat strabismus, but this is still in development.

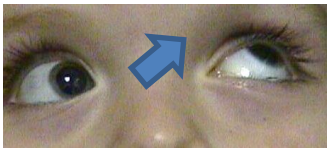


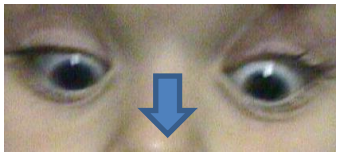
Patient in the photo right shows a left hypotropia that occurred 3 weeks after bupivacaine retrobulbar injection for cataract surgery. The left IR was inadvertently injected causing hypertrophy and increased muscle strength. Since the hypertrophic IR muscle overacts the hypotropia increases in downgaze.

**Question:** The child in the composite photos below has a 5 PD LHT in primary position, a significant chin elevation, and excellent horizontal and vertical saccades. There is no diplopia. Which is true?









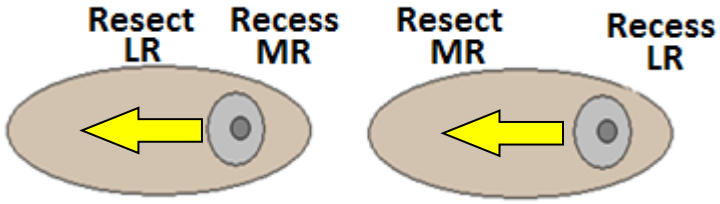
1. Right Knapp procedure best option
2. Will need full time occlusion for amblyopia
3. Wright Superior oblique silicone expander is indicated.
4. Recession of right inferior rectus is indicated.

**Answer:** There is no diplopia which suggests the patient can suppress and the strabismus is congenital or early childhood onset. The right eye has limitation of elevation much more in adduction than abduction suggestive of Brown's syndrome. On up-gaze there is a large exotropia so this is a V (or Y) pattern also consistent with Brown's syndrome. The treatment for Brown's syndrome is lengthening the SO tendon so the answer is # 3- Wright superior oblique silicone expander. What, did you really think I was going to end this book with someone else's procedure?

## Nystagmus with gaze preference – Face Turn

Patients with nystagmus will often have an eccentric gaze preference to place the eyes where the nystagmus is less and vision improves, called the null point. These patients will consequently adopt a face turn to keep their eyes at the null point. The child in the photo below has albinism with nystagmus and a left gaze preference (left null point). The left gaze preference causes a right face turn. The face turn can be improved by using strabismus surgery to move both eyes into primary position, i.e., **Kestenbaum procedure**. For the child in the photo below we would surgically move the eyes to the right, thus moving the eyes into primary position (see yellow arrows in drawing below). Perform a R & R each eye: right MR recession – LR resection, and left MR resection and LR recession. Parks showed that the amount of surgery needs to be huge and his suggested numbers are in the table below for a face turn of 30 degrees. These large surgical numbers will limit eye movements, therefore creating a new null point in primary position. Surgery is usually performed after 5 years of age but can be done earlier for extreme face turn.

Surgery for nystagmus with R face turn:




Left gaze preference so move the eyes to the right (to primary)

| | | LEFT EYE | | RIGHT EYE | |
|---|---|---|---|---|---|
| | Degree of Face Turn | Recess LR | Resect MR | Recess MR | Resect LR |
| **Parks** | 30° | 9 mm | 8 mm | 6.5 mm | 10 mm |

**Question:** Patient with motor nystagmus and chin elevation. Which is the best surgical option?

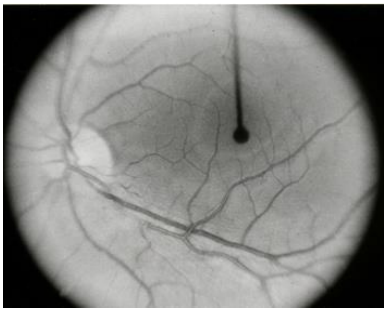
1. Recess IR OU and plication of SR OU
2. Plication if IR OU
3. Knapp transposition OU
4. IO recession with anteriorization
5. Recession SR OU

**Answer:** Compensatory chin elevation means the null point is in down gaze causing a down gaze preference. Perform a vertical Kestenbaum by weakening the IR muscles and tightening the SR muscles so the answer #1. This brings the eyes up into primary position.

# Addendum I Comprehensive Review Test and Answer Key

**CHAPTER 1: ANATOMY OF EYE MOVEMENTS**

## Question 1



Fundus photo direct view right shows:

**1.** Intorsion
2. Extorsion
3. Normal – No torsion
4. Pseudo-torsion

## Question 2

Which rectus muscle has the longest arc of contact to the globe?

1. Medial rectus
2. Lateral rectus
3. Inferior oblique
4. Superior oblique

## Question 3:

When hooking the medial rectus muscle in surgery it is necessary to pass the hook posterior to the limbus by at least:

1. 3.5 mm
2. 4.5 mm
3. 5.5 mm
4. 7.0 mm

## Question 4:

If lost during surgery which rectus muscle is most difficult to find?

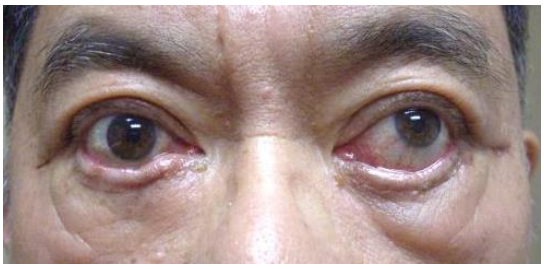
1. SR
2. IR
3. MR
4. LR

## Question 5:

When do the vertical rectus muscles have a single vertical function?

1. The eye adducts 23 degrees
2. The eye abducts 23 degrees
3. In downgaze
4. The vertical rectus muscles always have secondary and tertiary functions.

## Question 6:



Patient in photo right had bilateral lower lid blepharoplasty. Which best describes why the left eye is up and out, and the lower lid is retracted?

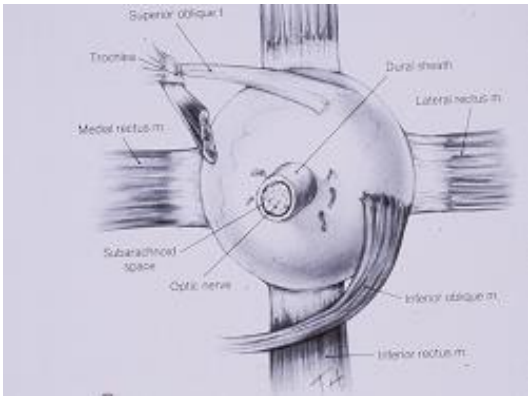
1. Secondary scarring of inferior orbital fat
2. Scarring of the lower lid skin incision
3. Traumatic disinsertion of the left IR muscle
4. Transection of SR muscle

## Question 7:

Which extraocular muscle group has the longest and shortest tendons?

1. Horizontal rectus muscles
2. Vertical rectus muscles
3. Oblique muscles
4. None of the above

## Question 8:



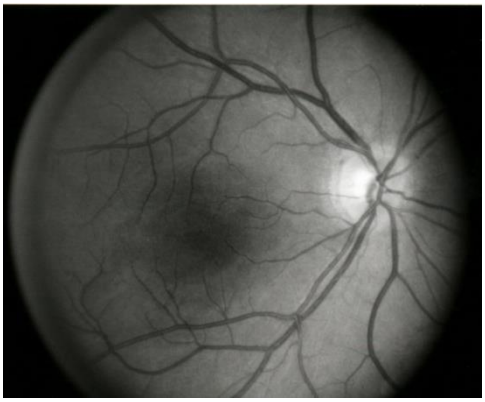

*Permission by T. Hengst, MA*

What is wrong with the drawing right from a popular anatomy textbook? (BTW - not one of my books)?

1. Inferior oblique muscle should be under the inferior rectus muscle.
2. SO insertion should extend posteriorly close to the optic nerve.
3. Both 1 and 2
4. It was published in a textbook so it has to be correct!

## Question 9:

Fundus photograph direct view of right eye in a patient with strabismus.

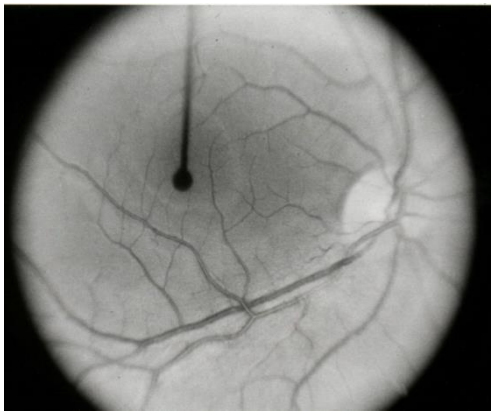


A. Which muscle could be weak?

1. SO
2. IO
3. IR
4. None of the above

B. Which surgical procedure would help correct this strabismus?

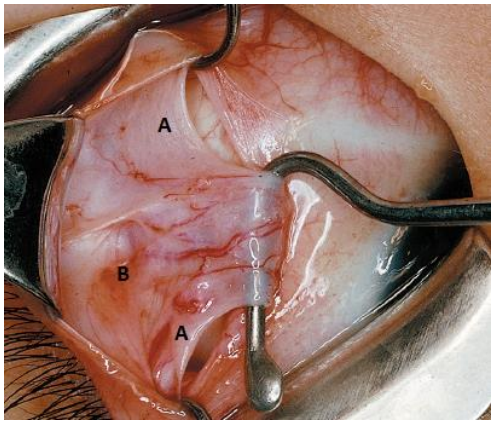
1. SR recession OD
2. IO recession OD
3. SO Harada Ito OD
4. # 2 and # 3



### Question 10:

Fundus photo direct view suggests which muscle or muscles are overacting?
(Could be more than one answer)

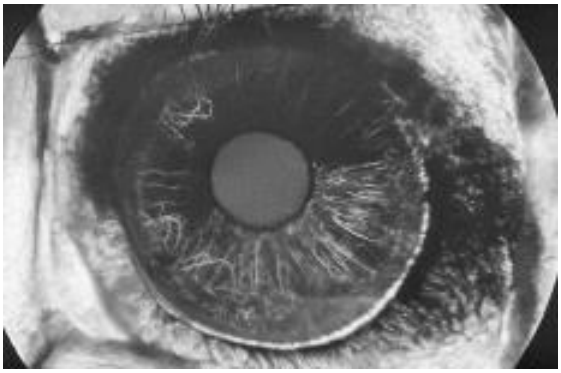
1. I.O.
2. S.O.
3. I.R.
4. S.R.




### Question 11:

The surgical photo right shows the lateral rectus muscle hooked and 2 small hooks under a white tissue above and below the lateral rectus muscle. Which of the tissues can be cut without causing the complication of fat adherence?

1. Tissue A
2. Tissue B
3. Both tissues A and B
4. None of the labeled tissues



### Question 12:

Photo right is an iris angiogram of the left eye taken after strabismus surgery that removed 3 rectus muscles. Which of the following describes the rectus muscle that was left intact.

1. Elevator, adductor and intorter
2. Depressor, adductor, and extorter
3. Pure abductor
4. Pure adductor

## Chapter 2 LAWS OF EYE MOVEMENTS

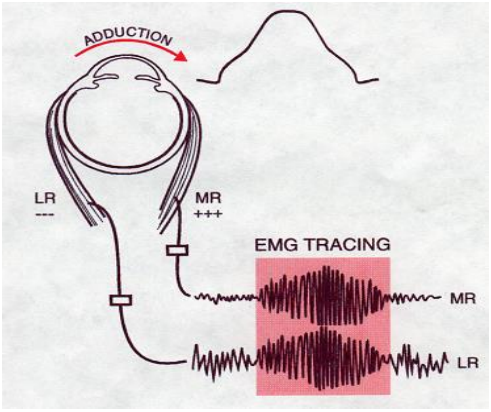
### Question 1:

The primary muscle action and field of action are in a similar direction for all of the following muscles except:

1. MR
2. IR
3. SR
4. SO

## Question 2:

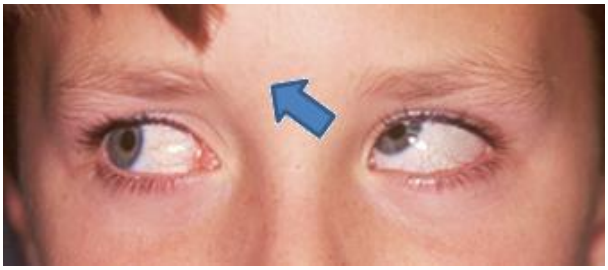
What is the best explanation for the EMG right?




1. Normal physiology of Sherrington's law
2. A violation of Sherrington's law caused by aberrant innervation with MR nerve splitting to innervate the MR and LR muscles, typical of Duane's syndrome.
3. The MR and LR show signs of neuromuscular disease
4. Violation of Listing's law of ocular rotation.

## Question 3:

Which muscle or muscles may be dysfunctional in this patient looking up and right?



1. Left IO over action
2. Left SO under action
3. Right SR weakness
4. Right IR restriction
5. All of the above

## Question 4:

Which of the following are true about convergence?

1. Corrects for our innate tendency for the eyes to diverge
2. Abides by Sherrington's Law
3. Violates Hering's Law
4. Strongest fusional vergence amplitude
5. All of the above

## Question 5:

A patient has an eye with limited abduction. The eye can generate an abduction saccade as the eye moves fast temporally, then stops abruptly. The limited abduction is caused by lateral rectus muscle palsy. **True or False?**

## Question 6:

A patient had a left medial rectus recession and developed a large exotropia immediately after surgery limited adduction left eye. The left eye adducts slowly, and there is no adduction saccade. Which of the following is true?

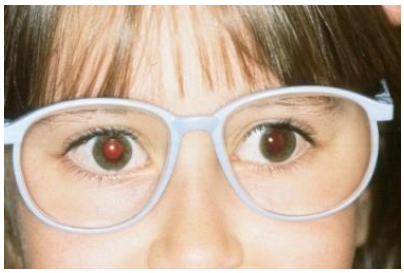
1. Rectus muscle with the shortest arc of contact is weak caused by the surgical complication of a lost muscle.
2. Left lateral rectus muscle is tight causing restriction and the left medial rectus function is good.
3. Surgical complication of scarring and restriction
4. #2 and #3 are correct

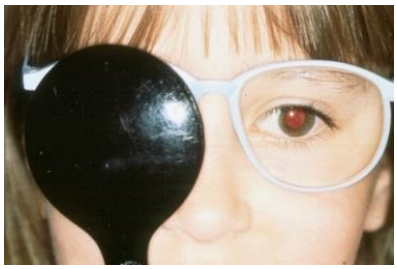
## Chapter 3 INTRODUCTION TO STRABISMUS

### Question 1:

Patient in photo shows good stereo acuity and an apparent left XT, however, covering right eye reveals no movement. The most likely diagnosis is:

1. Exotropia
2. Eccentric fixation
3. ARC
4. Convergence insufficiency
5. Orthotropia history of severe ROP



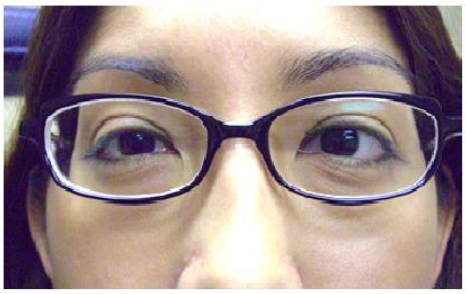


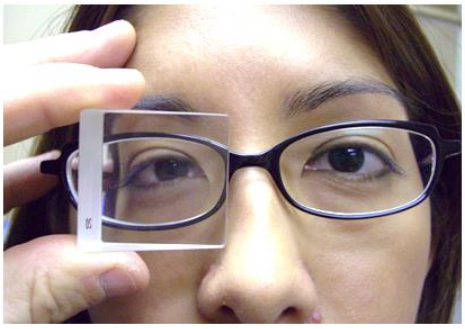
### Question 2:

Photo top right shows a patient with an esotropia fixing right eye because the left eye is blind. Bottom photo shows base out prism over the fixing right eye.

Which of the following is true regarding this patient?

1. Actually left eye is fixing
2. BO prism over the fixing right eye moves both eyes left per Hering's law
3. BO prism induces convergence
4. It is wrong to place the prism over fixing right eye for prism neutralization

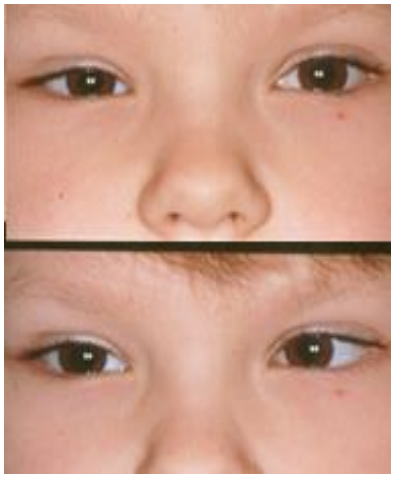




### Question 3:

If simultaneous prism cover test measures esodeviation 5 PD and prism alternate cover test measures esodeviation 20 PD the full esodeviation (tropia and phoria) is 20 PD. **True or False?**

## Question 4:



This 5 year old in photos right has an esotropia secondary to a post viral 6th nerve palsy. Note that the esotropia is greater when right eye is fixing (lower photo). All are true except:

1. Right 6th nerve palsy
2. Neutralize deviation with base out prism
3. Primary deviation is neutralized with prism over right eye
4. Secondary deviation is neutralized with prism over left eye
5. Limited abduction left eye

## Question 5:

Drawing (right) shows a patient fixing right eye and trying to looking up. The right eye has restricted elevation because the inferior rectus muscle is entrapped in a floor fracture. Which is correct?

1. Left hypertropia is comitant
2. Left hypertropia is incomitant and is greater in downgaze
3. Left hypertropia is incomitant and is greater in upgaze
4. Drawing shows the primary deviation

**Chapter 4 BINOCULAR VISION AND SENSORY ADAPTATIONS TO STRABISMUS**

## Question 1:

Why do we have a chiasm?

1. It gives us binocular vision and stereo acuity.
2. It gives us consensual pupillary response.
3. So we can be quizzed on Von Willebrand's knee
4. All of the above

## Question 2:

As a visual target moves to distance there is a reduction in stereo acuity. **True or False?**

## Question 3:

A patient complains of vertical diplopia so you place a red filter over the right eye. The patient sees the red image to be down, and tilted with the 12:00 o'clock position nasal. Which muscle is weak?

1. Right Inferior oblique
2. Left inferior oblique
3. Right superior oblique
4. Left superior oblique

### Question 4:

A five year old presents after having strabismus surgery at 8 months of age for congenital esotropia. Cover uncover test shows an esotropia 6 PD and alternate cover an esophoria of 12 PD. Worth four dot would show:

1. Three dots at distance and near
2. Four dots at distance and near
3. Three dots at distance and four dots at near
4. Four dots at distance and two at near

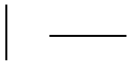
### Question 5:

This 40 year old has a long standing esotropia measured at 50 PD by prism alternate cover testing. What test result indicates if the patient has ARC and will likely see double if the esotropia is surgically corrected?

1. Prism neutralization with 50 base out (BO) causes diplopia
2. Synoptophore angled for ET 50 PD causes diplopia
3. Red filter test shows no diplopia
4. Answers # 1 and # 2

### Question 6:

An adult patient has an esotropia of 20 PD and prism neutralization with 20 PD BO gives the patient crossed diplopia. Afterimage test is given and the most likely response is:

1. A cross afterimage 
2. Lines of the afterimage side by side 
3. Vertical offset of the after image
4. Suppression of one of the afterimages

### Question 7:

An adult patient is complaining of intermittent horizontal diplopia. There is no shift on cover uncover testing and stereo acuity measures 40 seconds arc. What test or tests would best help make the diagnosis?

1. Bagolini lenses
2. Alternate cover test
3. Maddox rod
4. Answers #2 and #3

## Question 8:

A 40 year old with childhood strabismus develops a dense cataract after trauma to his dominant eye. After a year he now fixates with the previously non-dominant eye and the eye with the cataract is exotropic. Red filter test show crossed diplopia and diplopia persists with prism neutralization. You are planning cataract surgery. What should you tell the patient prior to cataract surgery?

1. No problem I'm an ace cataract surgeon.
2. Thank goodness you came to me rather than to those losers down the street.
3. Can you pay cash?
4. There is a good chance you will have persistent double vision and in some cases it does not go away even after strabismus surgery or prisms, and this is called "Horror Fusionis".

# Chapter 5 VISUAL DEVELOPMENT AND AMBLYOPIA

## Question 1:

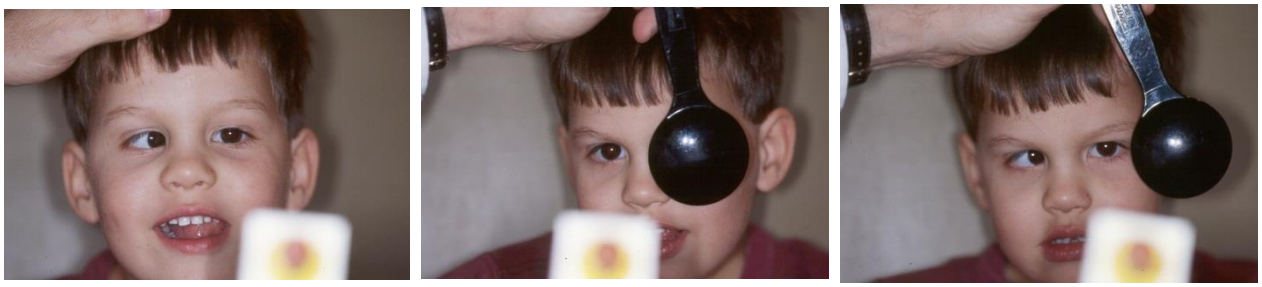
Which statement is true?

1. Amblyopia is always unilateral.
2. Amblyopia is "brain damage" in visual centers from abnormal visual stimulation.
3. Bilateral symmetrically blurred image in infancy does not cause amblyopia.
4. The critical period of visual development is the first year of life.

## Question 2:

Which of the following are correct for the child below?

1. No significant amblyopia
2. Mild amblyopia left eye
3. Significant amblyopia right eye
4. Cannot tell, let's wait a few years until the child is older and can read letters!



### Question 3:
Visual acuity difference between the good eye and amblyopic eye is greatest when vision is tested with:

1. Patient looking through a neutral density filter
2. Single letter presentation
3. Optotypes that have shape clues like the tree in Allen Cards
4. A line of optotypes presented to the patient

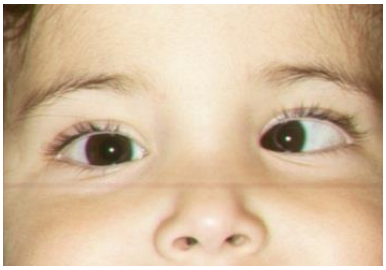
**Question 4:** A 3 year old presents with orthotropia and cycloplegic refraction of OD +1.00, and OS +4.00 sphere. You should immediately start full time patching of OD. **True or False**.

### Question 5:
A 5 year old presents with 20/80 OU and a cycloplegic refraction of + 8.00 OU. You should cut the plus and prescribe +4.00 OU – **True or False**?

### Question 6:
A 12 month old has a partial cataract right eye. Which is the best sign that the cataract is amblyogenic causing very poor vision?

1. Anterior location in lens
2. Orthotropia
3. Asymmetric red reflex
4. Eccentric fixation with poor following of a slowly moving object

## Chapter 6 ACCOMMODATIVE AND ACQUIRED ESOTROPIA




### Question 1:
The 3 yr. old in the photos right has a cycloplegic refraction of +3.00 OU, and the following deviation:

Nsc ET 40 D (Photo right top)
Ncc+3.00 OU E 10 (Photo right bottom)

Which of the following are true about the AC/A ratio?

1. Can't determine because the distance and near deviation is needed to determine the AC/A ratio.
2. The AC/A ratio is normal 5 PD/D.
3. The AC/A ratio is high 10 PD/D.
4. Cannot determine AC/A ratio because the inter-pupillary distance is required.

### Question 2:

At near a patient has an XT 25 then you give -2.00 lens OU in glasses. If the AC/A ratio is 5 PD/D (normal) what do you expect the deviation is with the myopic correction?

1. ET 5
2. Orthophoria
3. XT 15
4. XT 30

### Question 3:

A 5 year old with strabismus; at near (50 cm) an ET 10, at distance an XT 10, and the PD is 50 mm. The AC/A ratio by heterophoria formula: is 15 PD/D: **True or False.**

### Question 4:

The 4 year old is wearing hyperopic bifocal spectacles and for distance he is orthotropia. For near through the upper segment there is an ET 20 and lower segment orthotropia. Surgery is not indicated: **True of False**?

### Question 5:

A 3 year old presents with a recent onset intermittent esotropia measuring 30 PD for distance and near. Cycloplegic refraction shows +4.00 D. What would you do?

1. Prescribe full hyperopic correction - +4.00 D OU.
2. Reduce plus so patient will tolerate glasses – give +2.00 D OU.
3. Surgery – bilateral medial rectus recessions
4. Bifocal glasses

### Question: 6:

3 year old with an ET 20 PD distance and near after wearing +3.00 OU for 3 months. Multiple cycloplegic refractions verify a +3.00 sphere. Urgent surgery is indicated- **True or False?**

### Question 7:

A 3 year old presents with diplopia and an acquired intermittent esotropia 15 PD of two months duration. Dolls head maneuver shows limited abduction left eye. Cycloplegic refraction +1.00 OU and Fundus normal. Your management would be?

1. Prescribe glasses +1.00 OU then return in 6 months
2. Strabismus surgery to regain binocular fusion
3. Follow up in 6 months as strabismus often resolves
4. MRI of head

## Chapter 7 CONGENITAL ESOTROPIA

### Question 1:

A 40 year old presents to you with a history of previous strabismus surgery elsewhere for crossed eyes since birth. Your exam shows an alternating esotropia of 20 PD, nystagmus when one eye is occluded with fast phase to the fixing eye, R-hyper when RE covered and L-hyper when LE covered. Which one of the following is unlikely?

1. Bilateral DVD present
2. Patient has latent nystagmus
3. Monocular smooth pursuit asymmetry with poor pursuit nasal to temporal
4. Patient has amblyopia
5. Patient is stereo blind

### Question 2:

This 9 month old presents with an esotropia of 45 PD since birth. Patient alternates fixation, ductions are full and cycloplegic refraction is +1.00 OU. Which are most likely to be true?

1. DVD is present
2. Amblyopia present
3. Excellent prognosis for high grade stereo acuity.
4. Hyperopic glasses are indicated.
5. Esotropia will spontaneously resolve.
6. Subnormal stereo acuity - at best monofixation syndrome with peripheral fusion.

### Question 3:

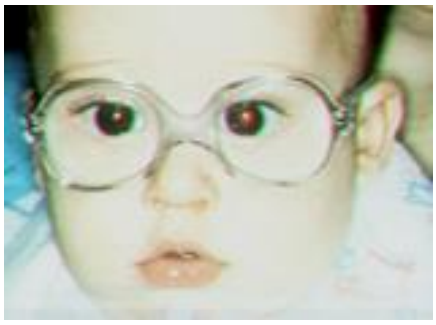
All of the following are true regarding infantile esotropia **except**:

1. Preoperative patching the dominant eye is indicated if there is strong fixation preference for one eye.
2. Esotropia is common in normal newborn infants.
3. Normal reading ability even though smooth pursuit asymmetry is present.
4. Surgery should be performed around 6 months of age to obtain peripheral fusion.

### Question 4:

After wearing full hyperopic correction +5.50 OU for 8 weeks the infant below (my son) has an ET 25 to 30 PD for distance and near with correction - full ductions. I was patching right eye 2 hours a day and he now holds fixation well each eye. What did I do next?

1. Obtain MRI scan of head
2. Increase patching right eye to 4 hours a day
3. Trial of bifocals
4. Urgent bilateral medial rectus recessions



## Chapter 8 EXOTROPIA

### Question 1:

Which of the following may cause the exotropia to become manifest in a patient with intermittent exotropia?

1. Covering one eye
2. Severely blurring vison of one eye
3. Taking a sedative like diazepam
4. Taking a few shots of whiskey
5. Attending a boring strabismus lecture by KW
6. All of the above

### Question 2:

Regarding intermittent exotropia which of the following is true?

1. Bifoveal fusion with high grade stereo acuity 40 sec. arc
2. Cortical suppression
3. Amblyopia rare
4. Onset usually after 2 years of age
5. All of the above

### Question 3:

10 year old with intermittent exotropia manifest most of the time. Cycloplegic refraction is plano OU. Deviation: Dsc X(T) 40 Nsc X(T) 20

The most appropriate treatment plan:

1. Convergence exercises
2. Prescribe glasses -3.00 sph OU
3. Bilateral lateral rectus recessions for XT 40
4. Bilateral lateral rectus recessions for XT 20
5. Perform patch test to determine surgical target angle

### Question 4:

Your 15 year old patient has a poorly controlled intermittent exotropia.
Deviation: Dsc X(T) 35 Nsc X(T) 10 Patch test = XT35

What do you want to do?

1. Patch for amblyopia
2. Prescribe bifocal spectacles
3. Bilateral lateral rectus recessions for XT 35
4. Bilateral lateral rectus recessions for XT 10

## Question 5:

An 8 year old presents with a history of having surgery elsewhere for intermittent exotropia 6 months ago. The child has diplopia at near since the surgery.
Your exam: Ductions full, Dsc Orthotropia, Nsc ET 20

Which is likely true regarding this case?

1. Patient has a high AC/A ratio.
2. Preoperative diagnosis was true divergence excess exotropia.
3. Bifocal glasses will help the diplopia
4. All the above

## Question 6

A 28 year old ophthalmology resident presents with poorly controlled intermittent exotropia and a chin elevation. Patching one eye eliminates the chin elevation. In addition to bilateral lateral rectus recessions which of the following is likely?

1. Need bilateral SO weakening procedure
2. Need IO muscle weakening OU
3. Need neck surgery
4. Chin up is because the patient is so proud that he/she understands the concept of ARC.

## Question 7:

Patients with which of the following would you expect to have equal vision and excellent stereo acuity of 40 sec?

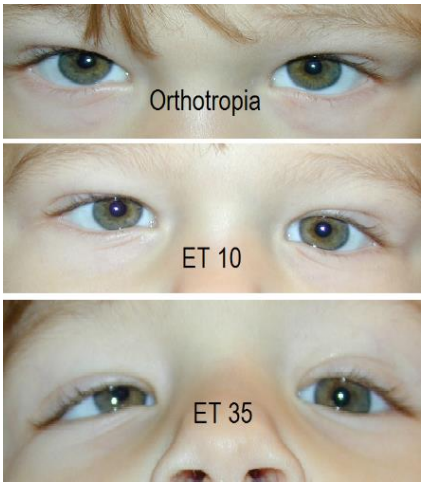
1. Infantile esotropia
2. Sensory exotropia
3. Intermittent exotropia
4. Congenital exotropia
5. None of the above

**Chapter 9 A AND V PATTERNS AND OBLIQUE MUSCLE DYSFUNCTION**

## Question 1:

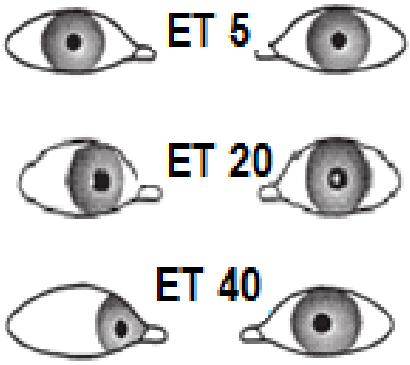
Pattern right is likely to have?

1. A pattern
2. Bilateral SO palsy
3. X pattern
4. Bilateral SO overaction



## Question 2:

A patient has an esotropia with the deviation shown right. There is no oblique dysfunction. What surgery is appropriate?




1. Bilateral medial rectus recessions with supraplacement
2. Bilateral medial rectus recessions with infraplacement
3. Bilateral lateral rectus resections with supraplacement
4. Bilateral medial rectus recessions without vertical transposition
5. Both 2 and 3

## Question 3:

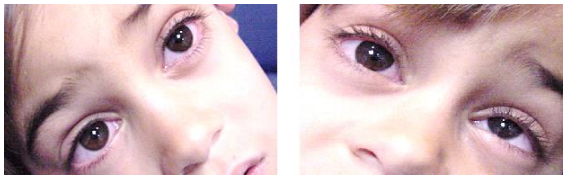
Patient in the composite photos below right has a RHT 10 in primary position.
What is the diagnosis?

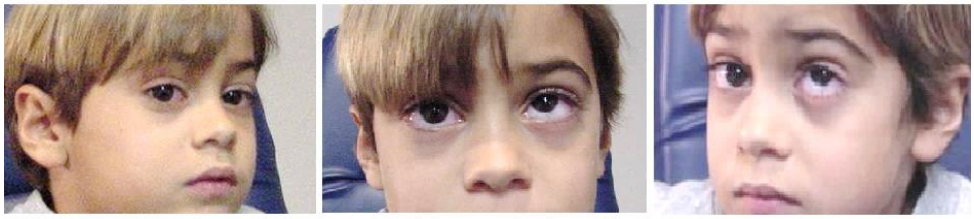
1. Right SR palsy
2. Right IR palsy
3. Right SO palsy
4. Left SO palsy
5. Left IO palsy


## Question 4:

Patient in photos below right has a LHT 10 in primary that increases to LHT 25 in left gaze.
Head tilt: Right LHT 2, Left LHT 18
Which muscle is paretic?

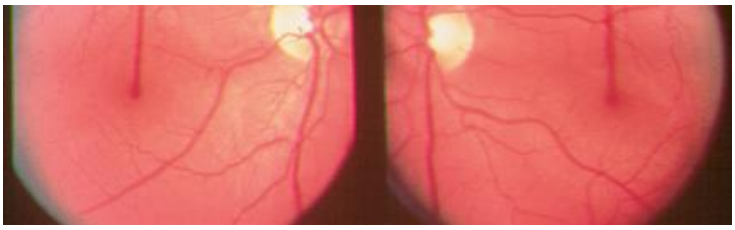
1. Left IR muscle
2. Left SO muscle
3. Right IO muscle
4. Right SR muscle





## Question 5:

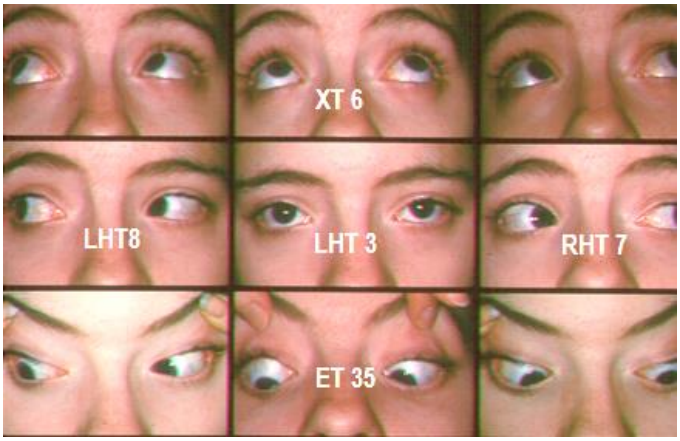
The fundus photos lower right are most consistent with which of the following?



1. Bilateral SO palsy
2. Bilateral IO over action
3. Masked bilateral SO palsy
4. All of the above

## Question 6:

The strabismus seen in the composite photos lower right: Head Tilt R- RHT 8, L- LHT10
This strabismus is most consistent with:




1. Bilateral inferior oblique palsy
2. Unilateral congenital SO palsy
3. Primary IO over action
4. Traumatic bilateral SO palsy

## Question 7:

The 8 year old patient had surgery for infantile esotropia at age 2 years. Now has vertical strabismus seen in composite photos right and no diplopia.
Primary: Orthotropia
Head Tilt: R - Ortho, L – Ortho
What is the diagnosis?

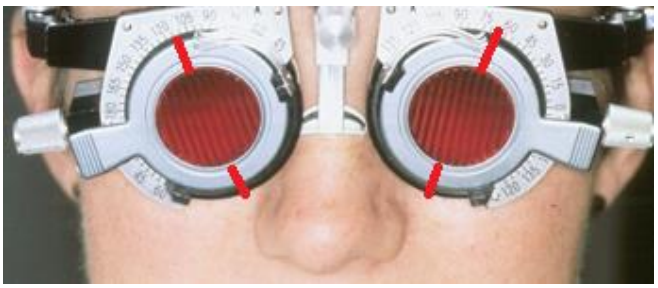
1. Bilateral SOP
2. Bilateral primary IOOA
3. R- SR palsy
4. Bilateral SR palsy

## Question 8:

What surgery could you do for the patient in question 7?

1. Bilateral inferior oblique recession
2. Bilateral inferior oblique myectomy
3. Bilateral inferior oblique anteriorization
4. All of the above

## Question 9:



Patient with a history of closed head trauma and torsional diplopia. Double Maddox rod is shown in photo right.
Primary - Orthotropia
Head Tilt; R- RHT 10, L- LHT 8
The most appropriate treatment is:

1. Prism glasses
2. Bilateral superior rectus recessions
3. Bilateral SO full tendon Tucks
4. Bilateral SO Harada Ito

## Question 10:

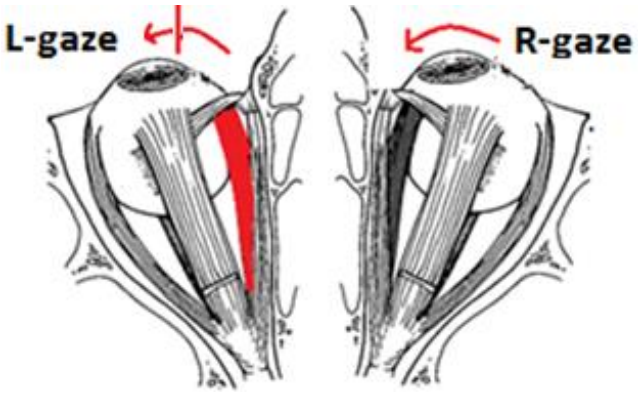
Patient in composite photos below has intermittent exotropia chin depression, and stereo acuity of 40 sec arc. Patient is bothered by the effort to fuse and wants strabismus surgery.
The preferred procedure is:

1. Bilateral LR recession
2. Bilateral LR recession and IO myectomy
3. Bilateral LR recession with supra-placement
4. Bilateral LR recessions and Wright SO tendon expander.

**Chapter 10 <u>RESTRICTIVE AND PARALYTIC STRABISMUS</u>**

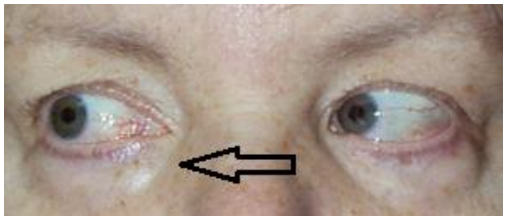
## Question 1:

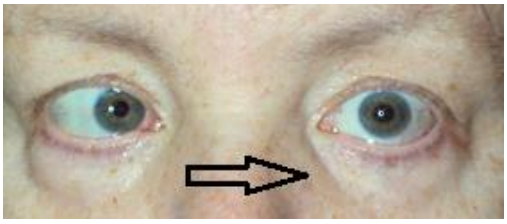
If the left MR muscle (in red) is tight:




1. The left eye shows limited abduction.
2. There is an esotropia increasing in left gaze.
3. Left lid fissure narrows with increased IOP in left gaze.
4. Left eye shows dog on a leash motility pattern in left gaze.
5. All the above

## Question 2:





The patient in the photos right can be best described as showing:

1. Restriction of the left eye to abduction
2. Paresis of the left lateral rectus muscle
3. Tight left medial rectus muscle
4. Limitation of abduction left eye
5. All of the above

## Question 3:

Patient in question 2 shows limited abduction left eye. How would you determine the lateral rectus muscle function?

1. Forced duction testing
2. Ability for the left eye to abduct past midline
3. Ability to generate an abduction saccade
4. Active force generation test
5. Both 3 and 4

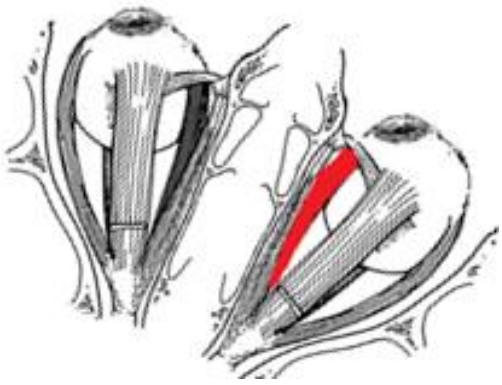
## Question 4:

Patient in question 2 with limited abduction shows a good abduction saccade before the eye stops abruptly. Patient cannot cooperate with forced duction testing. What is the cause of the limited abduction?

1. Left lateral rectus paresis
2. Restriction from a tight left medial rectus muscle
3. Paresis of the lateral rectus muscle and tight medial rectus muscle causing restriction
4. Cannot determine without forced duction testing

## Question 5:

Drawing depicts a face turn to the right and eyes left gaze. There is an esotropia in primary increasing right gaze, but the eyes are aligned in left gaze. The right eye has limited abduction but the abduction saccade is intact. Forced ductions indicate a tight right MR muscle (red). How would you correct the face turn?



1. Right medial rectus recession
2. Left lateral rectus muscle resection
3. Left lateral rectus muscle resection
4. Left medial rectus recession

## Question 6:

A patient has an acute traumatic bilateral 6th nerve paresis presents. What would you do?

1. Recession medial rectus OU
2. Ipsilateral MR recession and LR resection
3. Faden ipsilateral medial rectus
4. Wait at least 6 months before considering surgery.

## Question 7:

A traumatic left 6th nerve palsy occurred 6 months ago and the patient showed some recovery but now presents with diplopia and an ET 8 in primary which increases to ET 16 in left gaze. There is a good abduction saccade OU, but -1 abduction OS. Which is **FALSE**?

1. Uncrossed diplopia in left gaze
2. Residual mild left 6th paresis
3. Lateral rectus function is very poor
4. Recession of right medial rectus muscle would help the incomitance.

## Question 8:

A patient sustained a traumatic left 6th nerve palsy for 6 months and now presents with severe limitation of abduction OS and cannot generate an abduction saccade OS. One acceptable approach is:

1. Left Hummelsheim transposition and left MR recession
2. Right MR recession
3. Left MR recession
4. Left MR recession and LR Resection
5. Wait an additional year for 6th nerve recovery

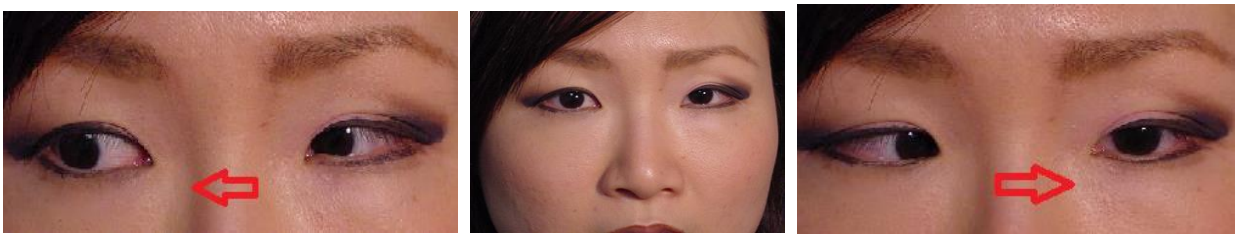
**Chapter 11 STRABISMUS SYNDROMES**

## Question 1:

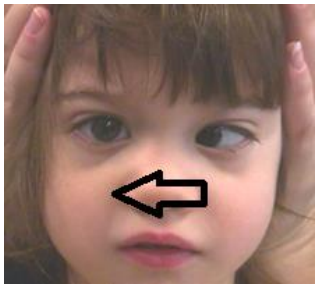
The patient below shows left eye has abduction deficit and lid fissure narrowing on adduction.

All are likely **except :**

1. Violation of Sherrington's law of agonist / antagonist
2. Abduction deficit left eye from agenesis of left abducens nucleus
3. Lid fissure changes are due to aberrant innervation of lateral by medial rectus nerve
4. Amblyopia LE



## Question 2:

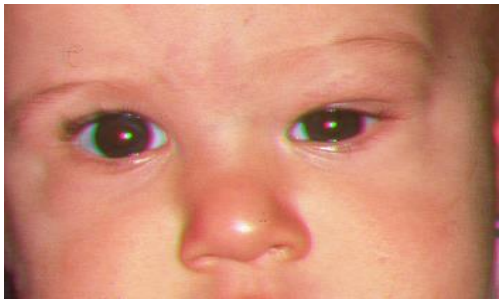


For the patient right which is correct?

1. Right type 1 esotropia Duane's syndrome
2. Gaze preference left with face turn right allows high-grade stereo acuity
3. Correct the face turn by recessing right MR muscle
4. Patient can suppresses diplopia yet has high grade stereo acuity
5. All of the above

## Question 3:

Infant in photo right has a swollen left upper lid and left hypotropia with limited elevation. What do you want to do?



1. Recess left IR muscle
2. Forced ductions at the time of surgery
3. Observe
4. MRI of orbit

## Question 4:

What is true about congenital Brown's syndrome?

1. Early surgery is critical.
2. Amblyopia is common.
3. Sensory adaptation like Duanes's syndrome and intermittent exotropia – high-grade stereo acuity and the ability for cortical suppression
4. Large hypotropia in primary position

## Question 5:

Congenital Brown's Syndrome is caused by an inelastic and tightness of the superior oblique muscle tendon complex as it passes through the trochlea- True of False?

## Question 6:

Patient in photo is 4 years old and presents with double vision and acquired limited elevation left eye. Best management is:

1) Release restriction by recessing left IR muscle
2) Knapp transposition
3) Recess right SR muscle to stop overshoot
4) MRI of orbit and brain

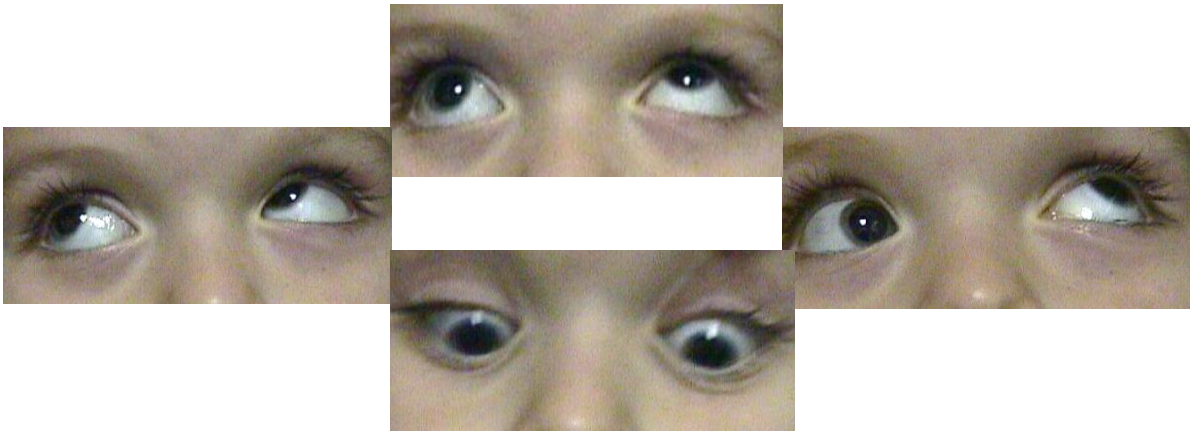
## Question 7:

8 year old shows poor elevation OS that is worse in abduction since infancy and no diplopia. There is a good up gaze saccade so you should recommend an IR recession: True or False?

## Question 8:

The child in the composite photos below has had abnormal eye movement since infancy. There is 5 PD LHT in primary position, orthotropia in down gaze, a significant chin elevation, and excellent horizontal and vertical saccades. There is no diplopia. Which is likely true?

1. Patient has binocular fusion
2. Will need full time occlusion for amblyopia
3. Diagnosis is MED or DEP
4. SR muscle palsy RE



## Question 9:

For the patient in question #8 above if you were to correct the chin elevation you would perform the Wright SO tendon expander; True or False?

## Question 10:

Patient with motor nystagmus and large chin elevation. Which is the best surgical option?

6. Recess IR muscles OU and plication of SR OU
7. Plication if IR muscles OU
8. Knapp transposition OU
9. IO muscle recession with anteriorization left eye
10. Recession SR muscles OU

# Answer Key

Ch 1
1. 1
2. 2
3. 4
4. 3
5. 2
6. 3
7. 3
8. 3
9. A 1, B 4
10. 2 and 4
11. 1
12. 2

Ch 2
1. 4
2. 2
3. 5
4. 5
5. False
6. 1

Ch 3
1. 5
2. 2
3. True
4. 5
5. 3

Ch 4
1. 4
2. True
3. 3
4. 3
5. 4
6. 2
7. 4
8. 4

Ch 5
1. 2
2. 3
3. 4
4. False
5. False
6. 4

Ch 6
1. 3
2. 3
3. True
4. True
5. 1
6. True
7. 4

Ch 7
1. 4
2. 6
3. 2
4. 4

Ch 8
1. 6
2. 5
3. 5
4. 3
5. 4
6. 2
7. 3

Ch 9
1. 2
2. 5
3. 3
4. 3
5. 4
6. 4
7. 2
8. 4
9. 4
10. 4

Ch 10
1. 5
2. 4
3. 5
4. 2
5. 1
6. 4
7. 3
8. 1

Ch 11
1. 4
2. 5
3. 4
4. 3
5. True
6. 4
7. True
8. 1
9. True
10. 1

# Addendum II Surgical Numbers

The following tables can be used as a guideline in planning strabismus surgery. These numbers have been derived from Parks, with modifications from the surgical experience of the author (Kenneth W. Wright, MD). The numbers are only a guide and should be modified as necessary.

## Binocular Surgery

**Esotropia**

| MR OU Recession | LR OU Resection* |
|---|---|
| 15$^{\Delta}$—3.0 mm | 15$^{\Delta}$—3.5 mm |
| 20$^{\Delta}$—3.5 mm | 20$^{\Delta}$—4.5 mm |
| 25$^{\Delta}$—4.0 mm | 25$^{\Delta}$—5.5 mm |
| 30$^{\Delta}$—4.5 mm | 30$^{\Delta}$—6.0 mm |
| 35$^{\Delta}$—5.0 mm | 35$^{\Delta}$—6.5 mm |
| 40$^{\Delta}$—5.5 mm | 40$^{\Delta}$—7.0 mm |
| 50$^{\Delta}$—6.0 mm | 50$^{\Delta}$—8.0 mm |
| 60$^{\Delta}$—6.5 mm | |
| 70$^{\Delta}$—7.0 mm | |

*When a lateral rectus resection is done for residual esotropia after a large medial rectus recession (6.0 mm or larger), these numbers should be lowered.

**Exotropia**

| LR OU Recession | MR OU Resection |
|---|---|
| 15$^{\Delta}$—4.0 mm | 15$^{\Delta}$—3.0 mm. |
| 20$^{\Delta}$—5.0 mm | 20$^{\Delta}$—4.0 mm |
| 25$^{\Delta}$—6.0 mm | 25$^{\Delta}$—5.0 mm |
| 30$^{\Delta}$—7.0 mm | 30$^{\Delta}$—5.5 mm |
| 35$^{\Delta}$—7.5 mm | 35$^{\Delta}$—6.0 mm |
| 40$^{\Delta}$—8.0 mm | 40$^{\Delta}$—6.5 mm |
| 50$^{\Delta}$—9.0 mm | |

Suggested surgical numbers by Kenneth W. Wright, MD

## Monocular Surgery

**Esotropia**

| MR Recession | LR Resection |
|---|---|
| 15Δ—3.0 mm | 3.5 mm |
| 20Δ—3.5 mm | 4.0 mm |
| 25Δ—4.0 mm | 5.0 mm |
| 30Δ—4.5 mm | 5.5 mm |
| 35Δ—5.0 mm | 6.0 mm |
| 40Δ—5.5 mm | 6.5 mm |
| 50Δ—6.0 mm | 7.0 mm |
| 60Δ—6.5 mm | 7.5 mm |
| 70Δ—7.0 mm | 8.0 mm |

**Exotropia**

| LR Recession | MR Resection |
|---|---|
| 15Δ—4.0 mm | 3.0 mm |
| 20Δ—5.0 mm | 4.0 mm |
| 25Δ—6.0 mm | 4.5 mm |
| 30Δ—6.5 mm | 5.0 mm |
| 35Δ—7.0 mm | 5.5 mm |
| 40Δ—7.5 mm | 6.0 mm |
| 50Δ—8.5 mm | 6.5 mm |

## Three Muscle Surgery

For large amounts of correction, surgery on three muscles may be planned for the primary operation. The amount of surgery can be judged from the above tables. This works especially well in adults, where one muscle can be placed on an adjustable suture. The adjustable suture should be done on the eye for which two muscles are being operated.

## Vertical Numbers

A rule of thumb for vertical surgery is 3 prism diopters of vertical correction for every millimeter of recession. Inferior rectus recessions are notorious for late overcorrections therefore, under most circumstances, do not recess the inferior rectus muscle more than 5 mm to 6 mm. Superior rectus recessions for dissociated vertical deviation (DVD), on the other hand, must be large, with the minimum recession of approximately 5 mm and a maximum of 9 mm (fixed suture technique).

Suggested surgical numbers by Kenneth W. Wright, MD

# Addendum III Wright Grooved Hook

For rectus muscle recessions, replace the standard large hook (e.g. Jameson hook) with the t itanium Wright grooved hook as seen in figure 1. The Wright grooved hook placed behind the muscle insertion allows for suturing the muscle insertion over the groove thus preventing inadvertent scleral perforation (figures 2-4). It also makes it easy to get full thickness locking bites, and keeps suture placement consistent: not too posterior – not too anterior. It is very helpful when suturing tight muscles as it pulls the muscle to the surgical field and provides a space to suture the muscle (figure 3). It is also useful for suturing the inferior oblique muscle insertion while protecting the sclera in the area of the macula. (Figure 4) Dr. Kenneth Wright holds a US patent on the hook design and it is sold at titansurgical.com.

Figure 1. Wright Groove Hook

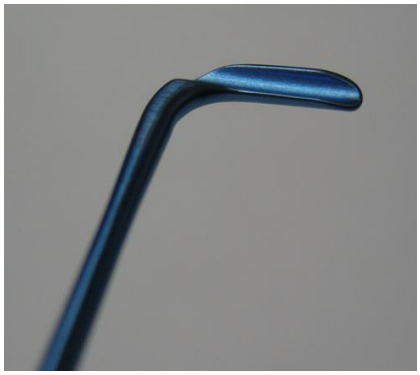


Figure 2. Drawing groove hook under rectus insertion

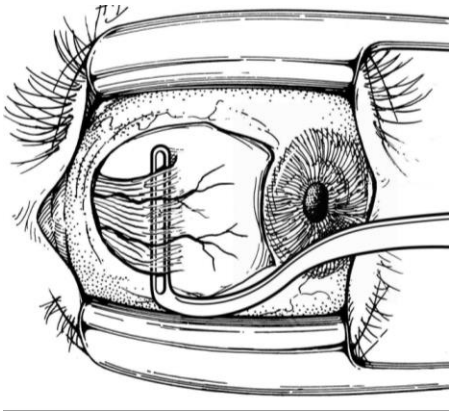


Figure 3. Suturing rectus muscle insertion

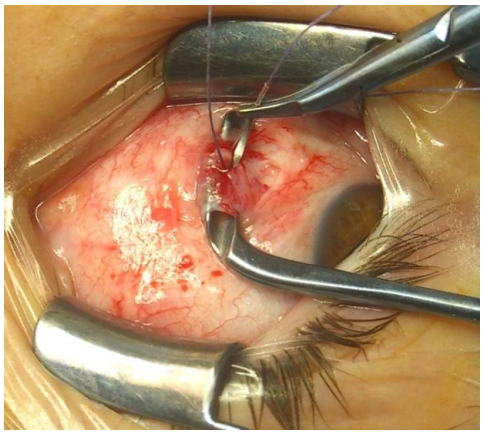
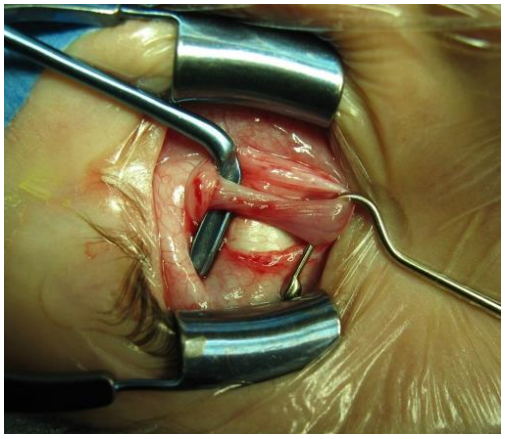


Figure 4. Groove hook under IO muscle insertion over groove hook



178 THE END