Grand Rounds

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Our Patient

- CC: “Right eye turns out sometimes”

- HPI: 4 year old Middle Eastern female presenting with intermittent episodes of right eye strabismus manifesting in vertical depression, often worsened by episodes of crying or stress as per mother. The patient has had a normal development and has no other visual complaints.

Core Competency: Patient Care

Our Patient

- PMH: full term, prenatal vitamins, normal development
- PSH: none
- POH: none
- Medications: none
- Ocular meds: none
- Allergies: none
- Family History: no blindness, glaucoma or strabismus
- Social History: none

Core Competency: Patient Care

Our Patient

Exam

- nVAsc: fixes and follows
- Dilated Refraction: plano OU
- Pupils 5-2 no APD OU
- EOM: see next slide
- CVF: unable because of age
- Tpalp: soft OU

Core Competency: Patient Care

Our Patient

Pen Light Exam

- LLA: wnl OU
- C/S: w/q OU
- K: cl OU
- I/P: r+r OU
- AC: f/s OU
- L: cl OU

Dilated Fundus Exam

- C/D 0.25 s+p OU
- M fl OU
- V wnl OU
- P no holes, tears, RD OU

Core Competency: Patient Care

Our Patient

Core Competency: Patient Care

Differential Diagnosis

Core Competency: Medical Knowledge

Differential Diagnosis

- Primary paralysis of inferior oblique
- Congenital fibrosis of inferior rectus muscle
- Endocrine orbitopathy
- Double elevator paralysis
- Fracture of orbital floor
- Brown Syndrome
- Pseudo-Brown Syndrome

Core Competency: Medical Knowledge

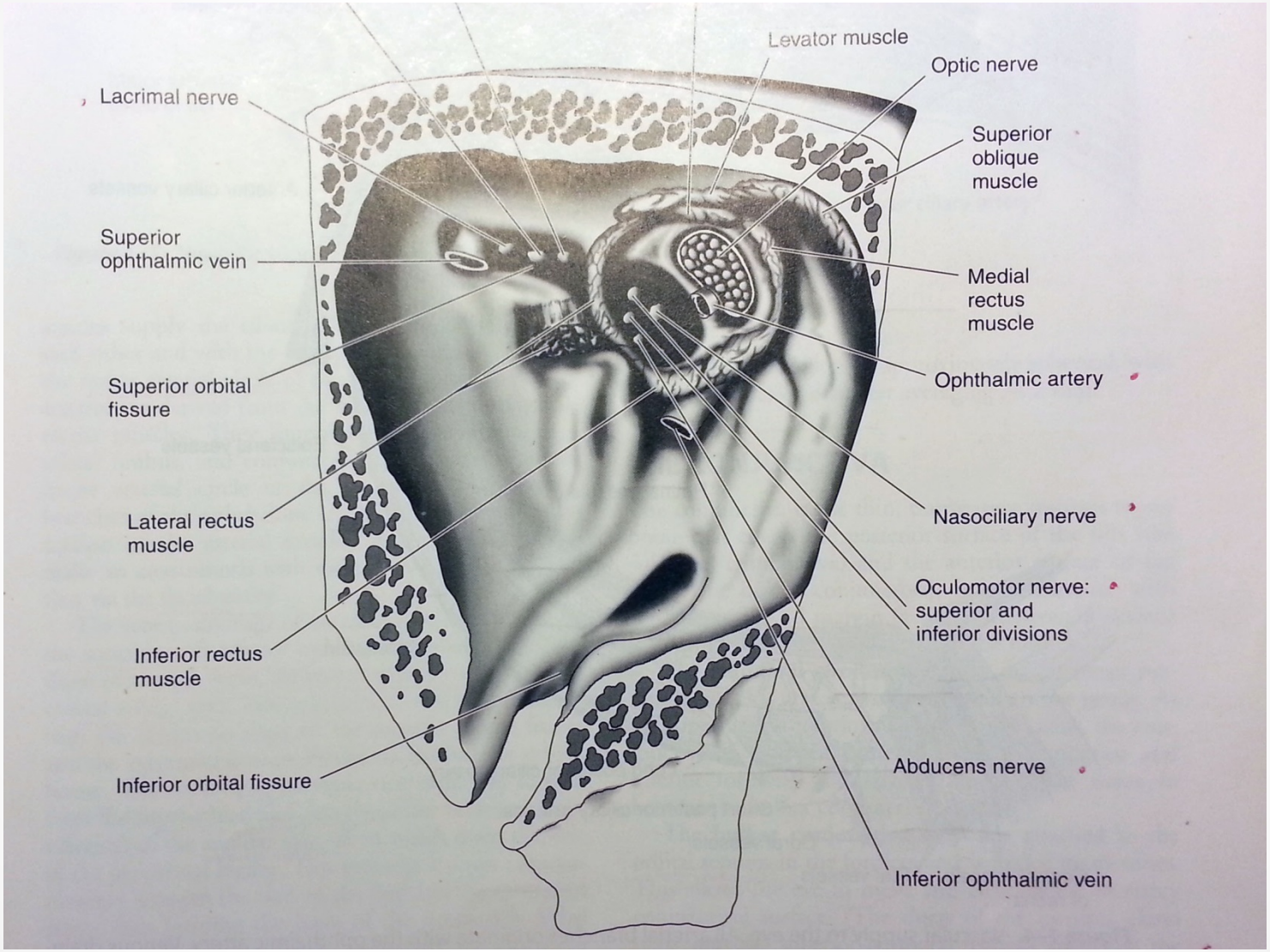


Image credit: Vaughan & Asbury's General Ophthalmology

Core Competency: Medical Knowledge

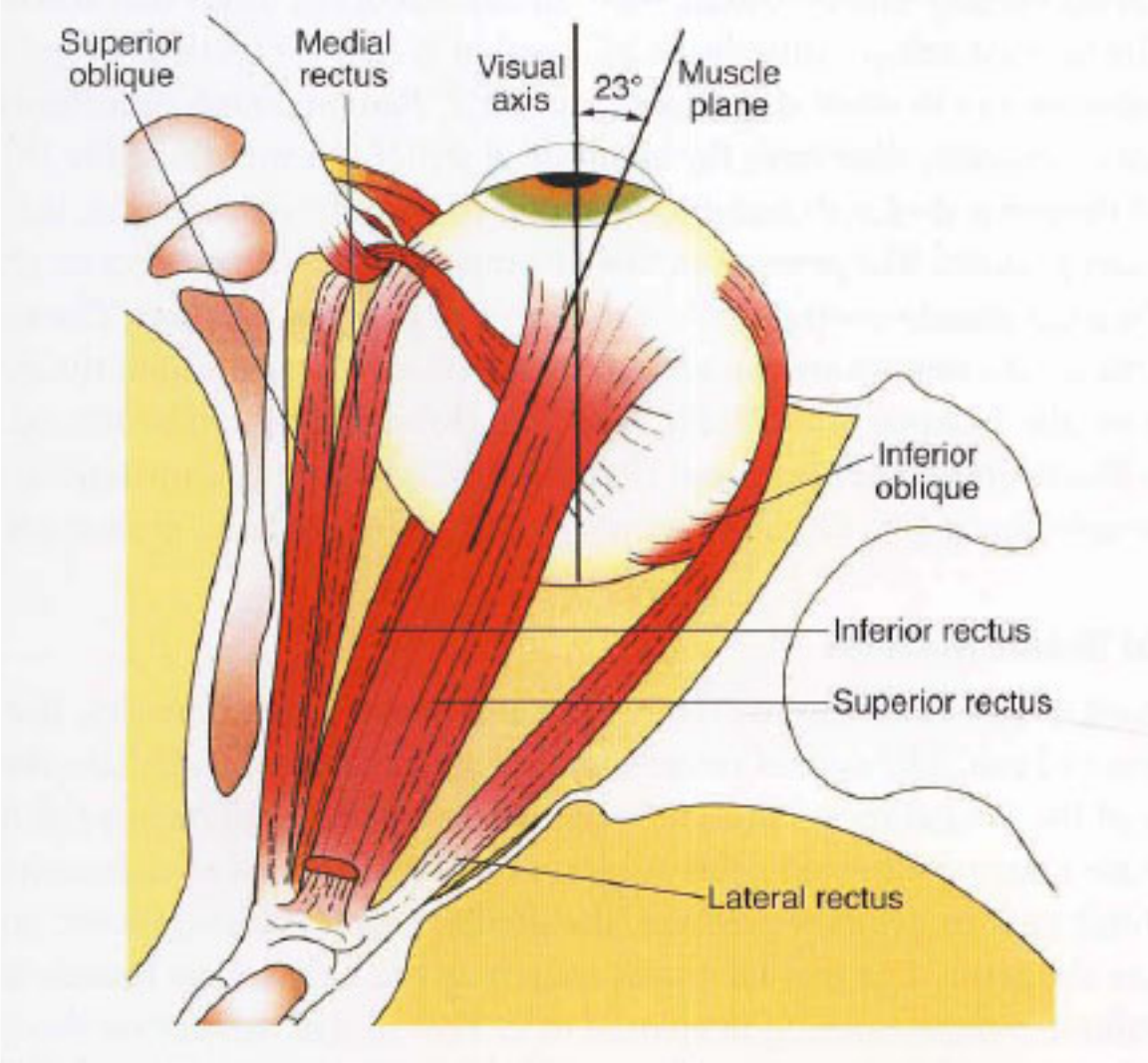


Image credit: BCSC

Core Competency: Medical Knowledge

Key Concepts

- Primary action of muscle
- Secondary and tertiary actions of muscle
- Maximum movement of globe
- Cardinal gaze positions
- Field of action
- Visual axis vs. muscle plane
- Superior oblique

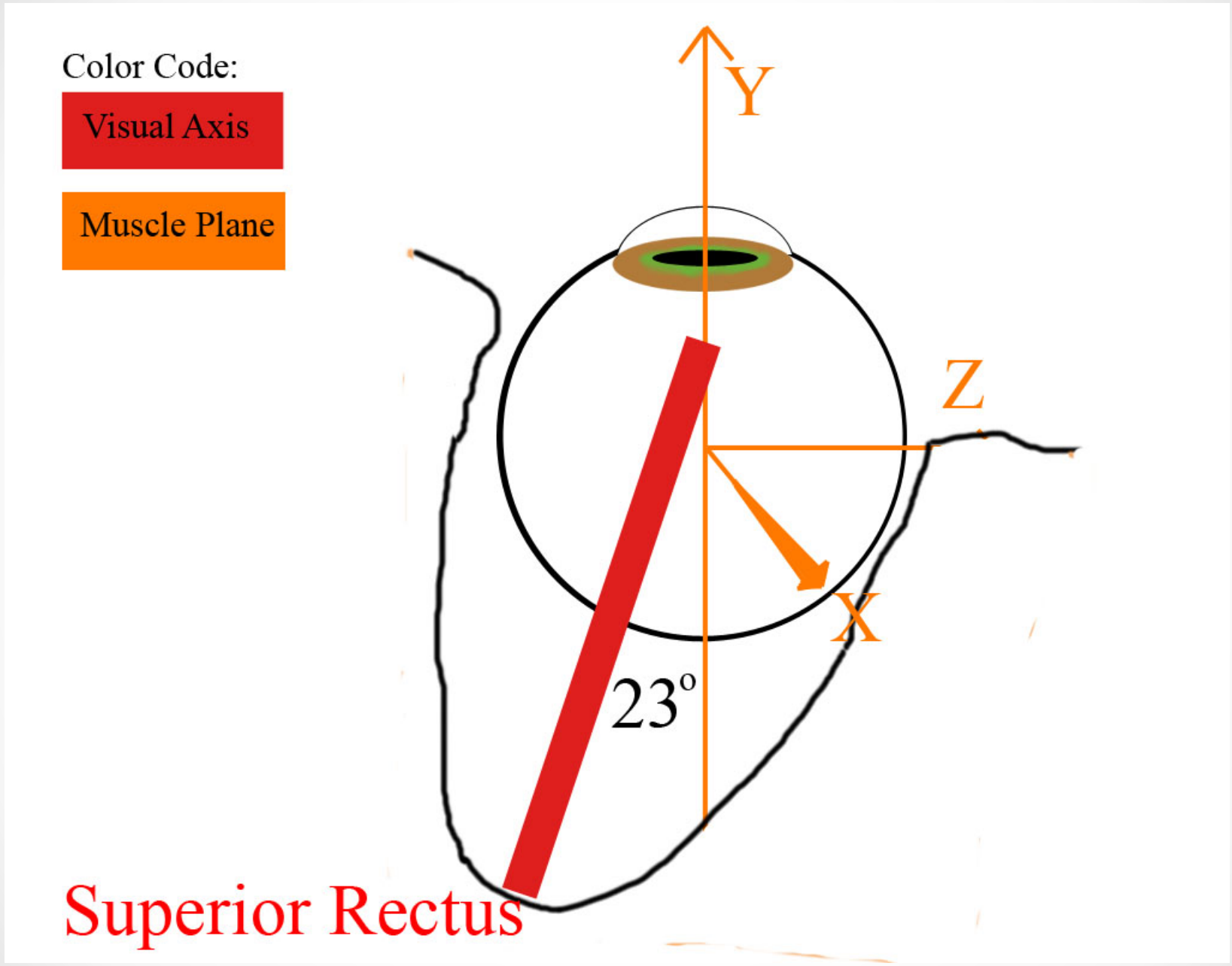
Core Competency: Medical Knowledge

Table 2-1 Extraocular Muscles

Muscle	Approx. Length of Active Muscle (mm)	Origin	Anatomical Insertion	Direction of Pull*	Tendon Length (mm)	Arc of Contact (mm)	Action From Primary Position	Innervation
Medial rectus (MR)	40	Annulus of Zinn	5.5 mm from medial limbus	90°	4.5	7	Adduction	Lower CN III
Lateral rectus (LR)	40	Annulus of Zinn	6.9 mm from lateral limbus	90°	7	12	Abduction	CN VI
Superior rectus (SR)	40	Annulus of Zinn	7.7 mm from superior limbus	23°	6	6.5	Elevation Intorsion Adduction	Upper CN III
Inferior rectus (IR)	40	Annulus of Zinn	6.5 mm from inferior limbus	23°	7	6.5	Depression Extorsion Adduction	Lower CN III
Superior oblique (SO)	32	Orbital apex above annulus of Zinn (functional origin at the trochlea)	Posterior to equator in superotemporal quadrant	51°	26	7–8	Intorsion Depression Abduction	CN IV
Inferior oblique (IO)	37	Behind inferior orbital rim lateral to lacrimal fossa	Macular area	51°	1	15	Extorsion Elevation Abduction	Lower CN III
Levator palpebrae superioris (LPS)	40	Orbital apex above annulus of Zinn	Septa of pretarsal orbicularis and anterior surface of tarsus	—	14–20	—	Eyelid elevation	Upper CN III

*Relative to visual axis in primary position. See also Chapter 3, Figs 3-3 through 3-7, in this volume.

Core Competency: Medical Knowledge

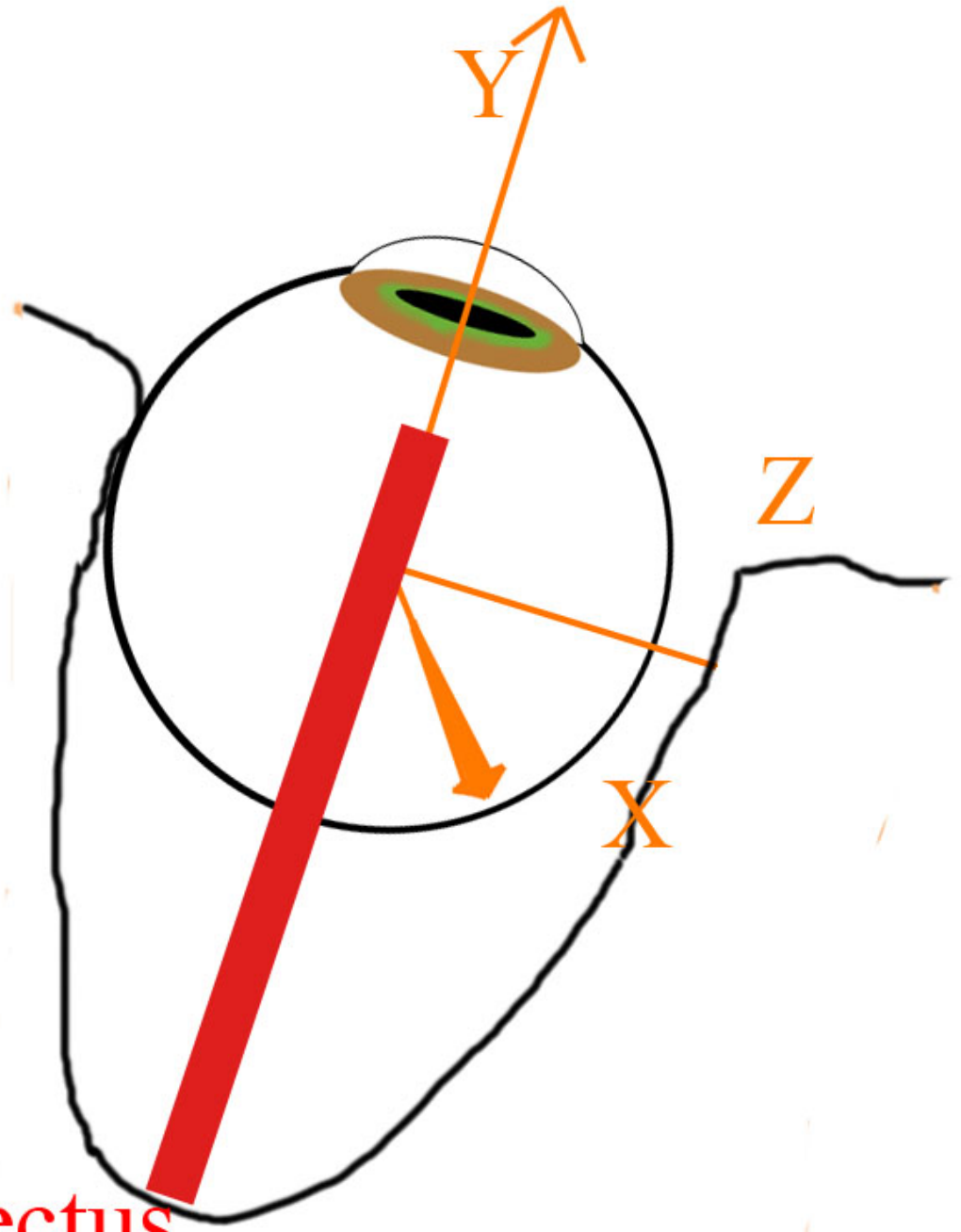


Core Competency: Medical Knowledge

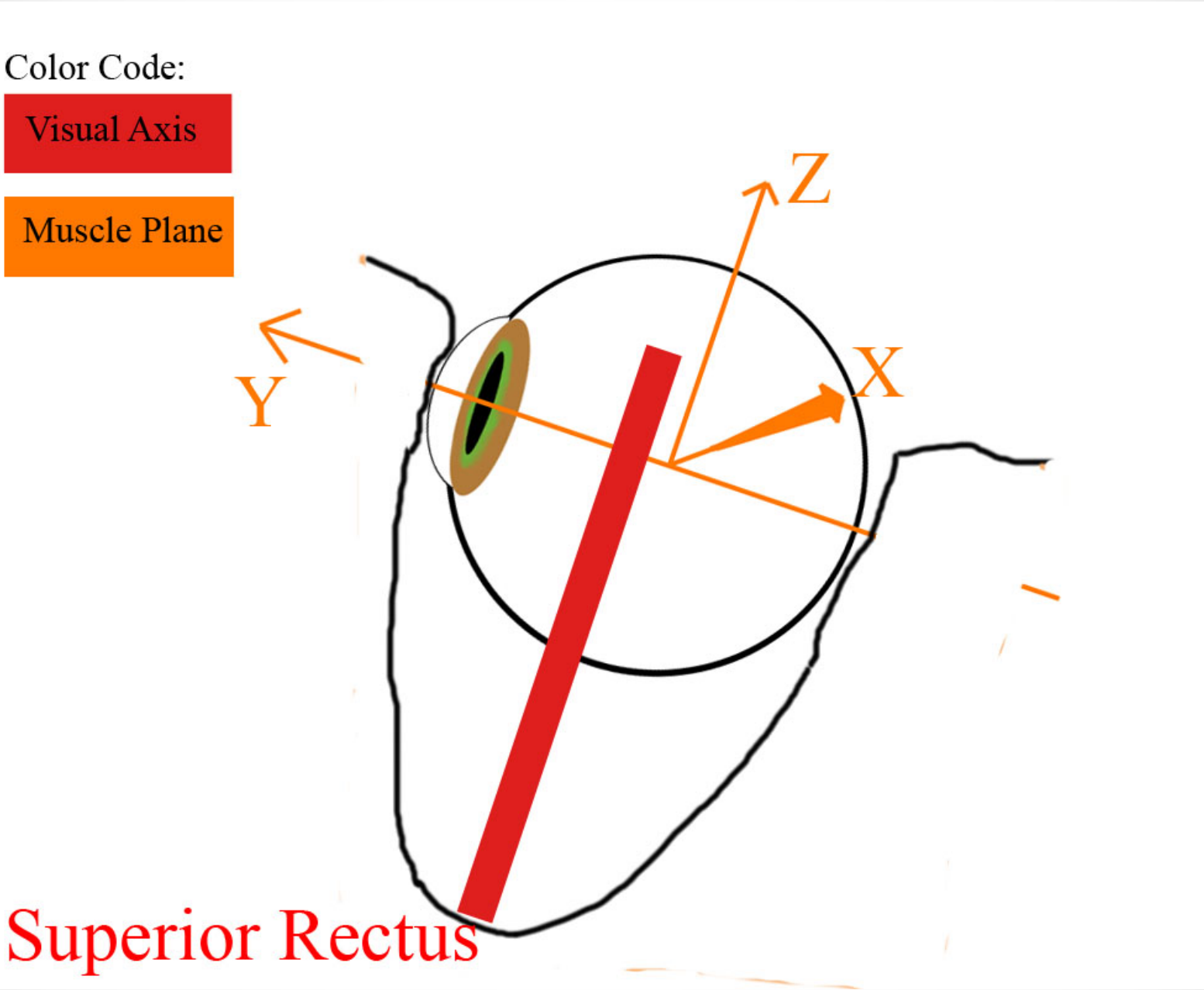
Color Code:

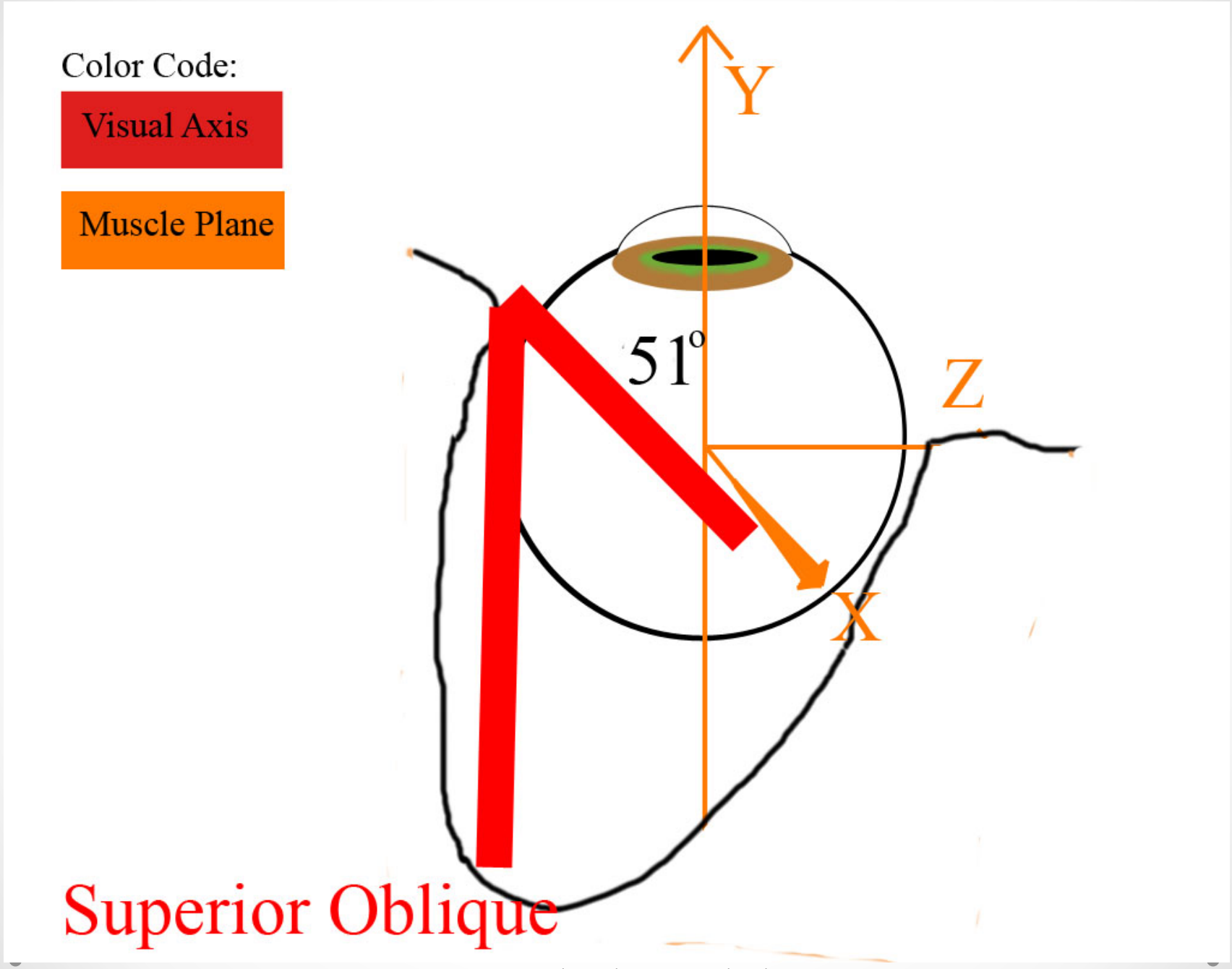
Visual Axis

Muscle Plane



Superior Rectus

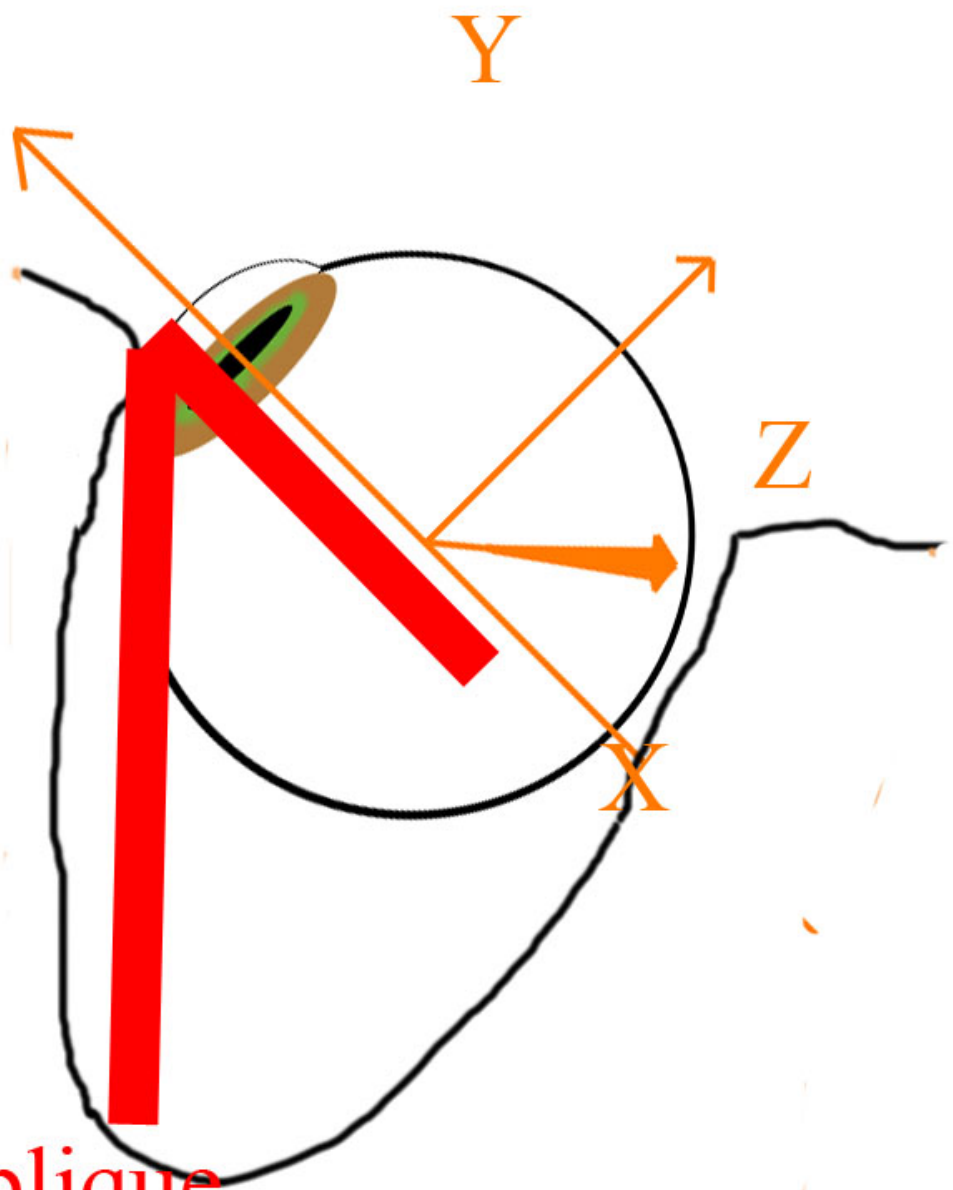

Color Code:
Visual Axis
Muscle Plane
Z
Y
X
Superior Rectus


Color Code:
Visual Axis
Muscle Plane
Y
51°
Z
X
Superior Oblique

Color Code:

Visual Axis

Muscle Plane

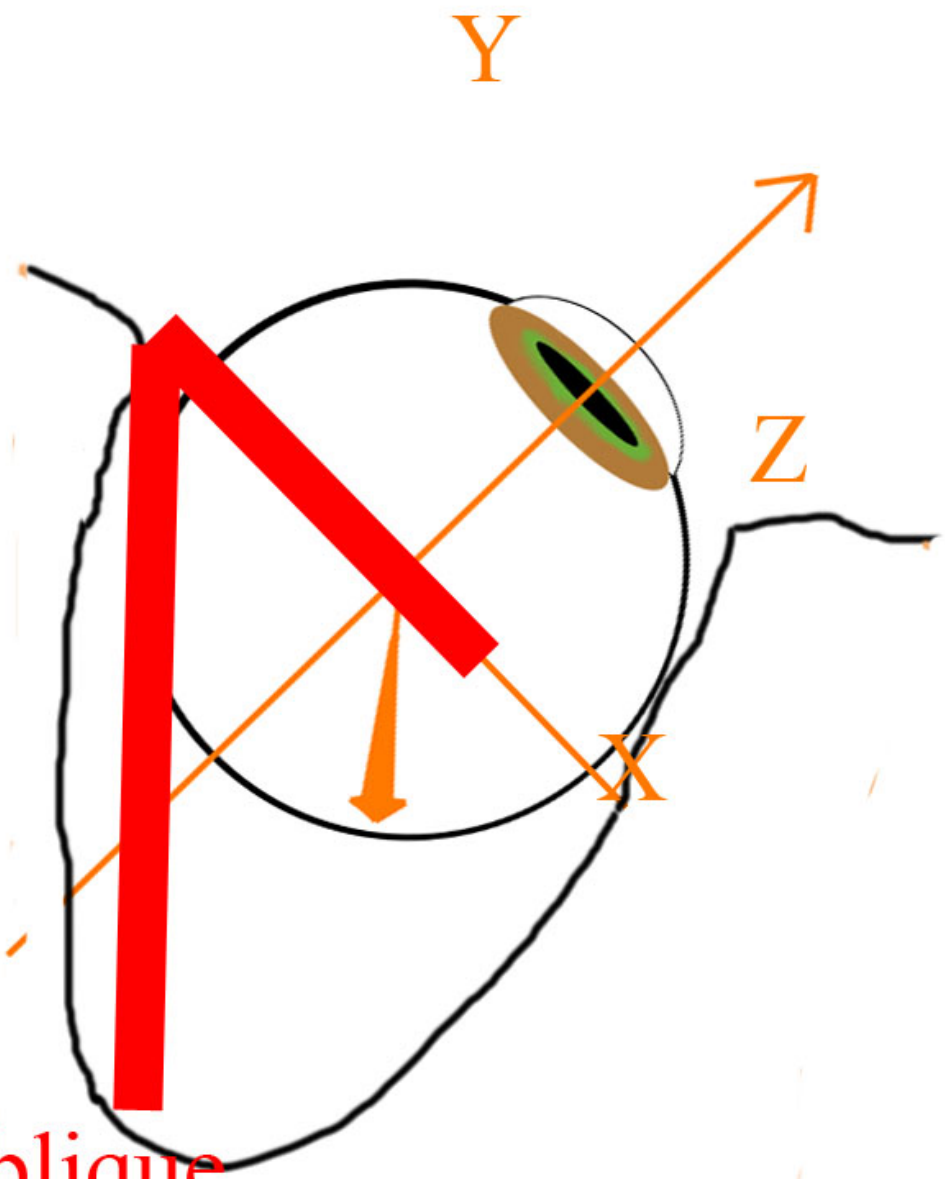


Superior Oblique

Color Code:

Visual Axis

Muscle Plane



Superior Oblique

Differential Diagnosis

- Primary paralysis of inferior oblique
- Congenital fibrosis of inferior rectus muscle
- Endocrine orbitopathy
- Double elevator paralysis
- Fracture of orbital floor
- Brown Syndrome
- Pseudo-Brown Syndrome

Core Competency: Medical Knowledge

Brown Syndrome

- Overview
  - Rare form of strabismus
- Mechanism
  - Sheath theory of superior oblique tendon (now debunked)
  - Now attributed to abnormalities of the tendon-trochlea complex leading to tight or short SO tendon

Core Competency: Medical Knowledge

Epidemiology

- Bilateral in 10% of cases
- 3:2 F:M and 2:1 OD:OS
- Congenital (more common)
- Acquired
  - Scleritis
  - Trauma to region of trochlea
  - Surgery (SO tucking / resection, blepharoplasty, sinus surgery, glaucoma drainage implant, scleral buckling)
  - Systemic inflammatory conditions near trochlea (tenosynovitis, RA, sarcoid, thyroid) leading to intermittent Brown Syndrome
  - Sinusitis leading to abscess near trochlea

Core Competency: Medical Knowledge

Symptoms and Findings

- Diploplia, pain
- Deficient elevation in adduction
- Improvement in abduction though often incomplete
- Widening of interpalpebral fissure in adduction
- Superior oblique click (click when restriction is released)
- Mild: no hypotropia in primary gaze
- Severe: hypotropia in primary gaze, chin up posture and face turn from affected side to maintain binocular fusion
- "Canine tooth" variant, described from dog-bite, combination Brown and SO palsy

Core Competency: Medical Knowledge

Diagnosis

- Forced duction in adduction: needs to be unequivocally positive
  - If restricted passive elevation is worsened in response to:
    - Retropulsion of globe (forceps-induced) → SO / Brown
    - Proptosis of globe (forceps-induced) → inferior rectus, etc
- V-pattern divergence on straight-ahead elevation
  - Not the case in IO paralysis
- Acute-onset of undetermined cause
  - Suspect sinusitis
  - CT orbits and paranasal sinuses

Core Competency: Medical Knowledge

Management

- Mild: observation: spontaneous improvement in 75% of cases in terms of "click", best resolution was -3 limitation to full movement, but complete resolution rare
  - When associated with RA / systemic inflammatory diseases, can use oral steroids or injected steroids to region of trochlea

Core Competency: Patient Care, Practice-Based Learning, Interpersonal and Communciation, Professionalism

Management

- Severe: surgery, may require more than one procedures, full motility restoration rare
  - Usually restricted to cases with compensatory head posture
  - Harold Brown sheathectomy
  - Z-myotomy
  - Ipsilateral superior oblique tenotomy
  - Iatrogenic SO palsy is frequent
    - Technique (careful handling of IMS, inert spacer, adjustable suture)
    - Simultaneous ipsilateral inferior oblique muscle weakening
  - Partial (80%) tenectomy of posterior tendon
  - Eye is anchored in elevated adducted position for two weeks after surgery

Core Competency: Medical Knowledge

Pseudo-Brown Syndrome

- Extraocular muscle involvement not including SO causing identical clinical picture as Brown Syndrome
- Lateral rectus pulley instability

Core Competency: Medical Knowledge

Reflective Practice

- This case represented application of eye muscle physiology and careful ophthalmic examination along with application of a focused differential diagnosis to evaluate an eye movement disorder.

Core Competency: Systems Based Practice

Core Competencies

Patient Care: The case involved thorough patient care and careful attention to the patient's past medical history.

Medical Knowledge: This presentation allowed me to review the presentation, differential diagnosis, proper evaluation/work up, treatment options for Brown Syndrome.

Practice-Based Learning and Improvement: This presentation included a literature search of current treatment modalities and background reading physiology.

Interpersonal and Communication Skills: The patient and her family were involved in the medical decision making

Professionalism: The patient provided information for the grand rounds presentation only after careful exam, explanation of findings and use of information.

Systems-Based Practice: The patient was managed by general and pediatric ophthalmology; general care for prenatal and developmental milestones provides information to make the correct diagnosis.

References

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- Albert and Jacobiec's Principles and Practice of Ophthalmology, 2008, p 4350, Elsevier Inc
- Wilson ME, Brown's Syndrome. Survey Ophthalmology 1989 Nov-Dec; 34(3): 153-72
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- Velez FG, Velex G, Thacker N. Superior oblique posterior tenectomy in patients with Brown syndrome with small deviations in the primary position. J AAPOS 2006:10(3): 214-219

Acknowledgements

- Patient
- Dr. Deutsch