

IOWA BASIC SCIENCE NOTES
Strabismus

2017

William E. Scott, M.D.

With contributions by
Scott A. Larson, MD

Department of Ophthalmology and Visual Sciences
University of Iowa

TABLE OF CONTENTS

<u>Visual Development in Children</u>	
Fixation Reflex	4
<u>Basic Examination Techniques for Children and Strabismic Adults</u>	
Basic Examination Techniques for Children and Strabismic Adults	6
Examining the Diplopic Patient	22
<u>Introduction to Strabismus</u>	
Incidence of Strabismus	24
Frequency of Types of Strabismus	24
<u>Esotropia</u>	
Definition	25
Etiology	25
Classification	26
Work-Up of a Patient with Esotropia	26
Pseudoesotropia	27
<u>Congenital Esotropia</u>	
Characteristics	28
Management of Congenital Esotropia	31
Duane's Syndrome	34
Moebius Syndrome	36
<u>Acquired Esotropia</u>	
VI Nerve Palsy	37
Accommodative Esotropia	40
Mydriatics, Cycloplegics and Anticholinesterases	43
Non-accommodative Acquired Esotropia	45
Prism Adaptation	46
PA for Near Angle	49
Esotropia Secondary to a Monocular Visual Defect	50
Factors Complicating the Course and Treatment of Esotropia	50
<u>Monofixation Syndrome</u>	
Characteristics	51
Features	51
Etiology	52
Tests for Monocular Scotoma During Binocular Viewing	52
Specific Characteristics	52
Treatment	53
Prognosis	54
<u>Exotropia</u>	
Classification	55
Characteristics	55
Etiology	55
Exophoria	56
Intermittent Exotropia	57
Congenital Exotropia	58

TABLE OF CONTENTS

(Continued)

Management of Exotropia

Non-Surgical Treatment	60
Surgical Treatment	61

Patterns Associated with Strabismus

Types of Patterns	64
Etiology	64
Principles of Treatment	66
Horizontal Muscle Shifts	66
Treatment of V-Pattern Esotropia	66
Treatment of V-Pattern Exotropia	67
Treatment of A-Pattern Esotropia	67
Treatment of A-Pattern Exotropia	67
Expected Correction from Oblique Muscle Weakening	69
Vertical Transposition of the Horizontal Rectus Muscles	69

Vertical Deviations

Differential Diagnosis	71
Superior Oblique Palsy	71
Monocular Elevation Deficiency	74
Inferior Oblique Palsy	76
Brown's Superior Oblique Tendon Sheath Syndrome	77
Blow-Out Fracture	79
III Nerve Palsy	81
Congenital Fibrosis Syndrome	84
Grave's Ophthalmopathy	84
Chronic Progressive External Ophthalmoplegia	89
Myasthenia Gravis	89
Cyclic Third Nerve Palsy	91

Nystagmus

Spasmus Nutans	93
Pendular Nystagmus	95
Latent Nystagmus	95
Congenital Nystagmus	97
Retrospective Review	99

Abnormal Head Positions Secondary to Motility Disorders

Abnormal Head Positions Secondary to Motility Disorders	103
---	-----

Supplement I - Amblyopia-Treatment & Results.....106

Visual Development in Children

I. Fixation Reflex

A. Characteristics

1. Requires structurally normal eye.
2. Ensures maximum visual acuity by projecting image onto fovea.
3. Optomotor reflex (involuntary)
4. May be interrupted by stimuli from:
 - a. Frontal lobe
 - b. Vestibular pathways
 - c. Auditory and pain pathways.

B. Visual developmental milestones

1. Premature
 - a. 30 weeks-blink to light
 - b. 31 weeks-pupil reactive to light
2. Neonate
 - a. 3-5 weeks-22% fixate face.
 - b. 9 weeks-90% fixate face.
 - c. 3 months-visually directed reaching.
 - d. 5 months-reaching/grasping

C. Estimated visual acuity of different ages

Age	Optokinetic Nystagmus	Forced Preferential Looking	Visually evoked potentials
Birth	20/400	20/400	20/100-20/200
2 months	20/400	20/200	20/80
4 months	20/200	20/200	20/80
6 months		20/150	20/20- 20/40
1 year	20/60	20/50	20/40
Age at 20/20	20-30 Months	18-24 Months	6-12 Months

(Most sensitive developmental age is the first 3 mos. of life-irreversible vision loss can occur if the vision is interrupted at this age)

D. Interference with fixation reflex development

1. Poor visual acuity, pendular nystagmus develops at age 3-6 months.
2. Congenital pendular nystagmus results in poor visual acuity.

E. Signs of reduced vision in infants and young children.

1. Unresponsiveness to visual stimuli.
2. Pendular nystagmus.
3. Strabismus.
4. Oculodigital reflex.

Basic Examination Techniques for Children and Strabismic Adults

I. Basic Examination Techniques for Children and Strabismic Adults

A. History

1. Ocular motility disorder
 - a. Age and mode of onset?
 - b. Which eye?
 - c. Which direction is misalignment?
 - d. Duration?
 - e. Documented photographically?
2. Description of symptoms
 - a. Blurring, asthenopia, diplopia?
 - b. Constant, intermittent, variable?
 - c. Worse in gaze?
 - d. Distance or near?
 - e. Monocular or binocular?
3. Known cause associated with onset
4. Previous treatment
 - a. Glasses: When first prescribed? How current is prescription? Improvement in condition?
 - b. Prisms.
 - c. Occlusion: Which eye? Period of time?
 - d. Exercises: Description
 - e. Drugs: What type?
 - f. Surgical: Which eye? How many muscles? How many times?
5. Family history
 - a. Taking a careful family history including several generations will often reveal esotropia and exotropia if that is the presenting symptom
6. Birth history
 - a. Prematurity
7. General health/growth and development? Medications? Diagnosed syndromes?

B. Sensory exam

1. Worth 4-Dot
 - a. Central @ distance (6m)
 - b. Peripheral @ near ($\frac{1}{3}$ m)

2. Polarized 4-Dot

*Arthur BW, Marshall A, McGillivray D. Worth vs. polarized four-dot test. J Ped Ophthalmol Strab 1993; 30:53-55.

3. Stereotests

- a. Titmus.
- b. Randot.
- c. Distance Vectograph.
 - i. new projection machines may not offer this feature

C. Measuring the basic deviation

1. Alignment as measured with proper spectacle correction.
2. Accommodation is controlled with an accommodative target.
 - a. Scott fixation device.
 - b. Finger puppets.
 - c. Wiggle sticks.
 - d. Snellen acuity chart.
 - e. Video.



Scott Fixation Devices

3. Fusion is suspended.
 - a. 45-minute occlusion is used for intermittent deviations but not necessary in constant deviations because fusion is suspended.

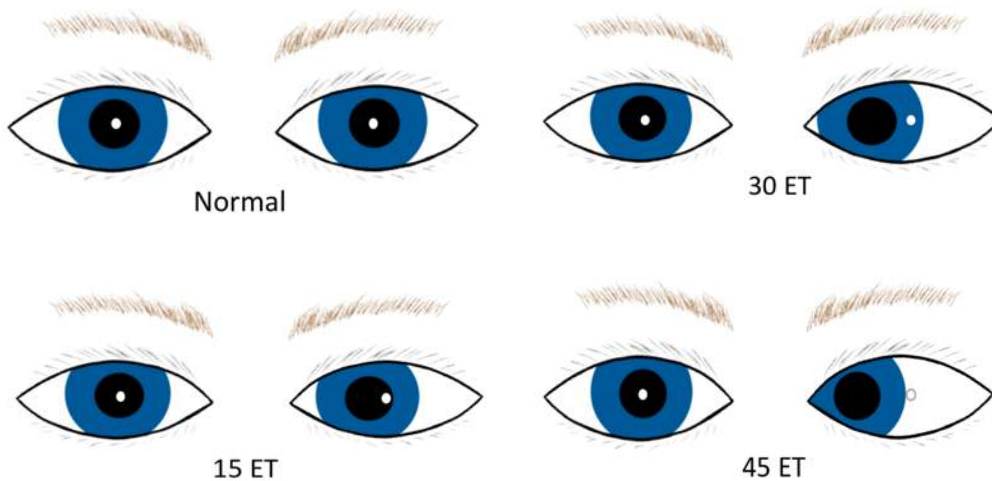
*Scott WE, Mash AJ, Redmond MR. Comparison of accommodative and non-accommodative targets for the assessment of ocular deviations. Amer Orthop J 1976; 26:83-86.

D. Detection of strabismus

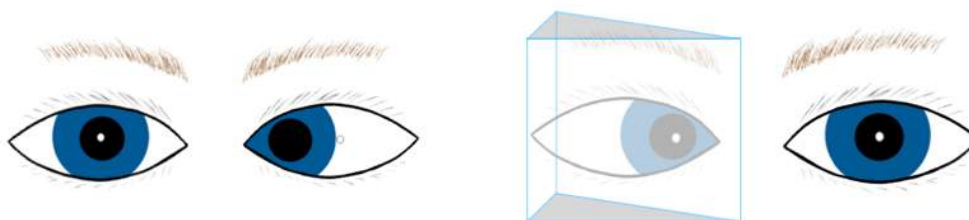
1. Single Cover Test (SCT) and Cover/Uncover Test.
 - a. Fusion is not suspended.
 - b. Detects presence of tropia.
2. Alternate Cover Test (ACT)
 - a. Detects presence of phoria.
 - b. Detects basic deviation.
3. Cover testing video demonstration on EyeRounds.org
<http://webeye.ophth.uiowa.edu/eyeforum/video/basic-cover-test.htm>

E. Quantitation of strabismus

1. Corneal Reflection Tests
 - a. Hirschberg - corneal light reflex.



- b. Modified Krimsky - prism held in front of fixing eye.



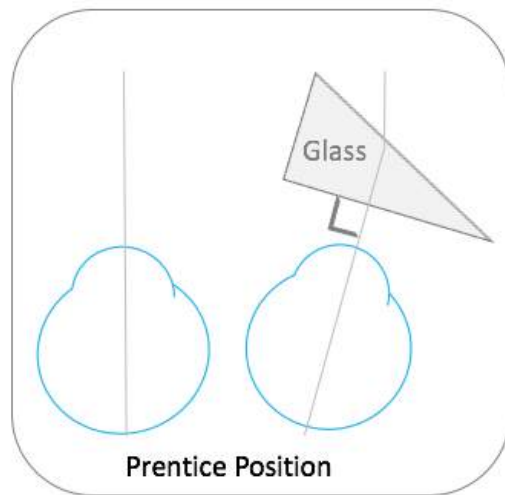
- c. Accommodation is not controlled.
 - d. Fusion is not suspended.
2. Prism Cover Tests
 - a. Single prism cover test.
 - b. Simultaneous cover and prism test.
 - c. Alternate cover and prism test.
 3. Scleral comparison
 4. Subjective detection

*Larson SA, Keech RV, Verdick RE. The Threshold for the Detection of Strabismus. J AAPOS. 2003; 8:418-422

F. Positioning of prisms

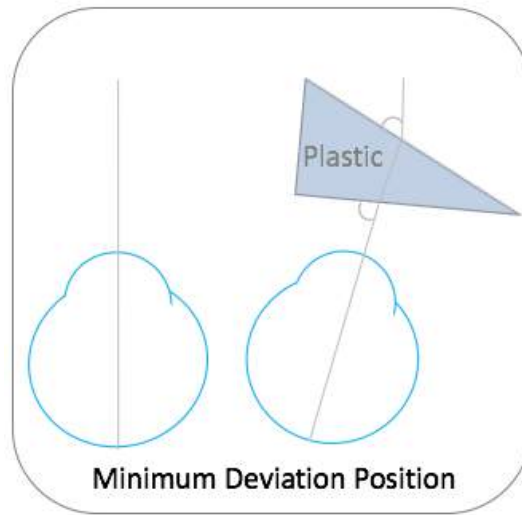
1. The deviation that a prism produces or neutralizes is dependent on the position of the prism as it is held before the patient. Remember--when prisms are used to measure a strabismus deviation, the prism displaces the image so that no movement of either eye is needed to fixate on the target.
 - a. Prentice position.

The line of sight of the deviated eye is perpendicular to the posterior face of the prism. This is the proper way to hold a glass prism.



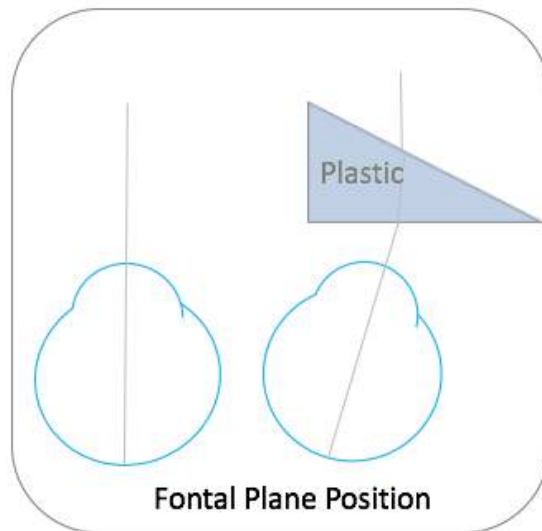
- b. Minimum deviation position.

The visual axis makes an equal angle with each prism surface. This is the position in which plastic prisms are calibrated.



c. Frontal plane position.

The posterior face of the prisms is held in the frontal plane of the patient. This is the most common way of holding plastic prisms and most closely approximates the minimum deviation position.



d. Horizontal and vertical prism bars are calibrated for use in the frontal plane position.

e. Fresnel press-on prisms are calibrated in the Prentice position.

2. Prism held perpendicular to the line of sight anterior to the prism when measuring lateral gaze, vertical gazes, or near.
3. Prism held with base parallel to the lateral wall or floor of the orbit when measuring in head tilt.

4. Never stack two prisms in the same direction as this causes large measurement error. Stacking a vertical and a horizontal prism together causes no measurement error.

Actual Prism Value of Two Prisms Stacked in the Same Direction

	1 st Prism				
2 nd Prism	10	20	30	40	50
10	21	33	46	60	77
20	33	47	66	89	122
30	46	66	94	141	264
40	60	89	141	339	
50	77	122	256		

5. Splitting prisms between the two eyes to measure large deviations causes some measurement error but substantially less than stacking.

- * Thompson JT, Guyton DL. Ophthalmic prisms. Measurement errors and how to minimize them. *Ophthalmol* 1983; 90(3):204-210.
- * Helveston EM. Prism placement. Measurements of horizontal and vertical deviations with the head tilted. *Arch Ophthalmol* 1975; 93:483-486.
- * Thompson JT, Guyton DL. Ophthalmic prisms. Deviant behavior at near. *Ophthalmol* 1985; 92(5):684-690.
- * Scattergood KD, Brown MH, Guyton DL. Artifacts introduced by spectacle lenses in the measurement of strabismic deviations. *Amer J Ophthalmol* 1983; 96(4):439-448.

G. Variables with the Prism Cover Test

1. Prisms
 - a. Positioning.
 - b. Stacking.
 - c. Splitting.
2. Spectacles
 - a. High plus or minus spectacles (generally ≥ 5 D) create a built in prismatic effect in strabismic patients that must be taken into account when performing a PCT.
 - b. High minus glasses will induce a base in prism effect for esotropes and a base out prism effect in exotropes. In both cases this will have the effect of making the deviation by the PCT appear larger than the true deviation by 2.5 D %. (i.e., a patient wearing -10.00 D glasses with a 40^Δ esotropia by PCT will have a true deviation of 30^Δ).
 - c. High plus glasses will induce a base out prism effect for esotropes and base in effect for exotropes. In both cases this will make the measured deviation smaller than the true deviation by 2.5 D % (i.e., a patient wearing +10.00 D glasses with a 40^Δ esotropia by PCT will have a true deviation of 50^Δ).

d. Anisometropic spectacles.

Prism is induced in the presence of an anisometropic spectacle correction when the visual axis is not aligned with the optical axis of the lenses. This can cause diplopia and account for differences in measurements obtained in secondary and tertiary gaze positions.

True Ocular Deviation Measured with Hyperopic Spectacles

	Spectacle Power													
Angle	+1	+2	+3	+4	+5	+6	+7	+8	+9	+10	+12	+15	+20	+30
5	5	5	5	6	6	6	6	6	6	7	7	8	10	20
10	10	11	11	11	11	12	12	13	13	13	14	16	20	40
15	15	16	16	16	17	18	18	19	19	20	21	24	30	60
20	21	21	22	22	23	24	24	25	26	27	29	32	40	80
25	26	26	27	28	29	30	31	32	33	35	37	40	50	100
30	31	32	32	33	34	35	36	38	39	40	43	48	60	120
35	36	37	38	39	40	41	42	44	45	47	50	56	70	140
40	41	42	43	44	46	47	48	50	52	53	57	64	80	160
45	46	47	49	50	51	53	55	56	58	60	64	72	90	180
50	51	53	54	56	57	59	61	63	65	67	71	80	100	200
60	62	63	65	67	69	71	73	75	77	80	87	96	120	240
70	72	74	76	78	80	82	85	88	90	93	100	112	140	280

True Ocular Deviation Measured with Myopic Spectacles

	Spectacle Power													
Angle	-1	-2	-3	-4	-5	-6	-7	-8	-9	-10	-12	-15	-20	-30
5	5	5	5	4	4	4	4	4	4	4	4	4	3	3
10	10	10	9	9	9	9	9	8	8	8	8	7	7	6
15	15	14	14	14	13	13	13	12	12	12	12	11	10	9
20	20	19	19	18	18	17	17	17	16	16	15	15	13	11
25	24	24	23	23	22	22	21	21	20	20	19	18	17	14
30	29	29	28	27	27	26	26	25	24	24	23	22	20	17
35	34	33	33	32	31	30	30	30	29	28	27	25	23	20
40	39	38	37	36	36	35	34	33	33	32	31	29	26	23
45	44	43	42	41	40	39	38	37	37	36	35	33	30	26
50	49	48	47	45	44	43	43	42	41	40	38	36	33	29
60	59	57	56	55	53	52	51	50	49	48	46	44	40	34
70	68	67	65	64	62	61	60	58	57	56	54	51	46	40

3. Incomitant deviations

- A & V patterns.
- Paresis.
- Restriction.
- Anomalous innervation.

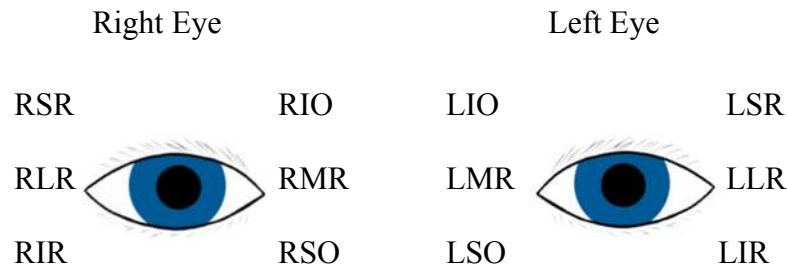
4. Variable deviations

- Cyclic deviations.
- Nystagmus blockage.
- DVD.
- Myasthenia.
- Thyroid.
- CNS disorder.

H. Examination of ocular movement (ductions & versions)

- Ductions: movement of a single eye in any direction.
- Versions: movement of both eyes in any direction.
- Tests are based on

- a. Hering's Law (of equal innervation): When innervation is sent to a muscle causing it to contract, equal innervation goes to its yoke muscle (contralateral synergist) in order to maintain parallelism of the visual axes.
- b. Sherrington's Law (of reciprocal innervation): When one extraocular muscle receives an impulse to contract, its ipsilateral opposing muscle (direct antagonist) receives an impulse to relax, allowing smooth movement to take place.
- c. Muscle responsible for primary movement from primary position



4. Synergists: Two or three muscles of the same eye that work together to produce a given movement.

<u>Action</u>	<u>Muscles</u>
Elevation	SR, IO
Depression	IR, SO
Adduction	MR, SR, IR
Abduction	LR, SO, IO
Extorsion	IO, IR
Intorsion	SO, SR

5. Yoke Muscles: Pairs of extraocular muscles that move both eyes together in a parallel movement in the nine directions of gaze.

<u>Gaze</u>	<u>Yoke Muscles</u>
Right	RLR, LMR
Left	RMR, LLR
Elevation	RSR, RIO, LSR, LIO
Depression	RIR, RSO, LIR, LSO
Up & Right	RSR, LIO
Down & Right	RIR, LSO
Up & Left	RIO, LSR
Down & Left	RSO, LIR

6. On examining ocular movement, the extent of movement in each direction of gaze, as well as the quality of that movement, should be noted.

The patient should be asked to fixate a target moving from primary into the limits of the eight positions of gaze. Versions should be tested first, then ductions.

- * Keep patient's head straight throughout the examination.
- * Test depression without holding lids up first to note associated anomalies of lid movement.
- * Test ductions in case of limitation on versions and manifest strabismus.

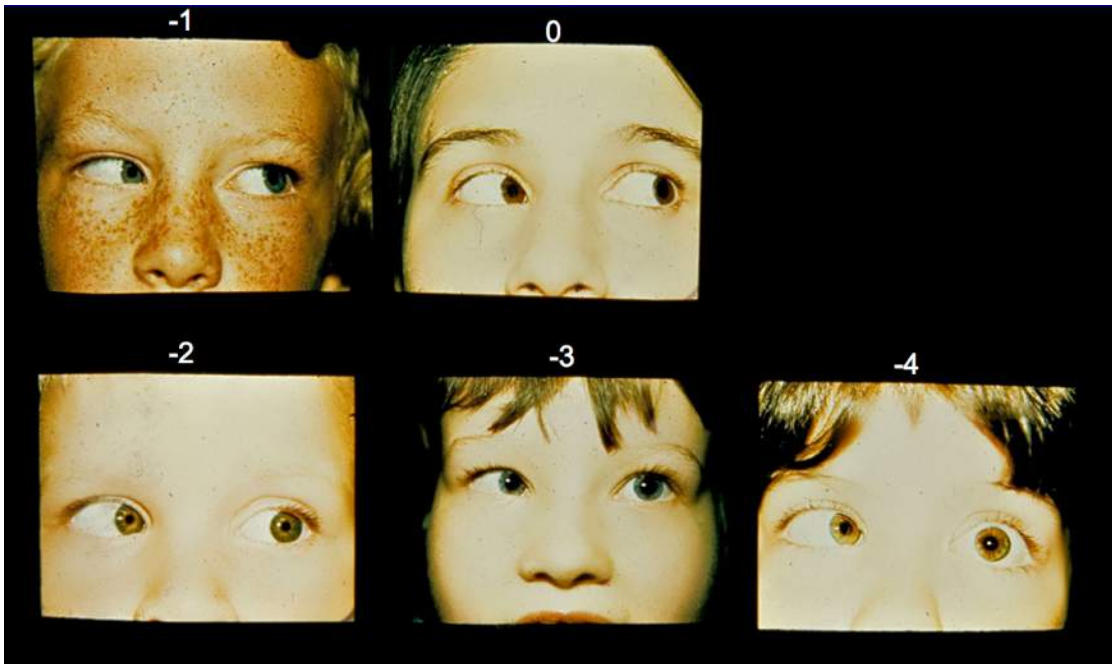
7. Observations

Abnormalities of ocular movement can be seen by comparing the amount of visible sclera, corneal reflections or the position of the limbus. Variations in palpebral fissure shape and size can give a misleading impression. The following observations should be noted and recorded.

- Underaction and overaction (-4 -> 0 -> +4).
- Grading of ductions from - 4 to + 4 with "0" being normal



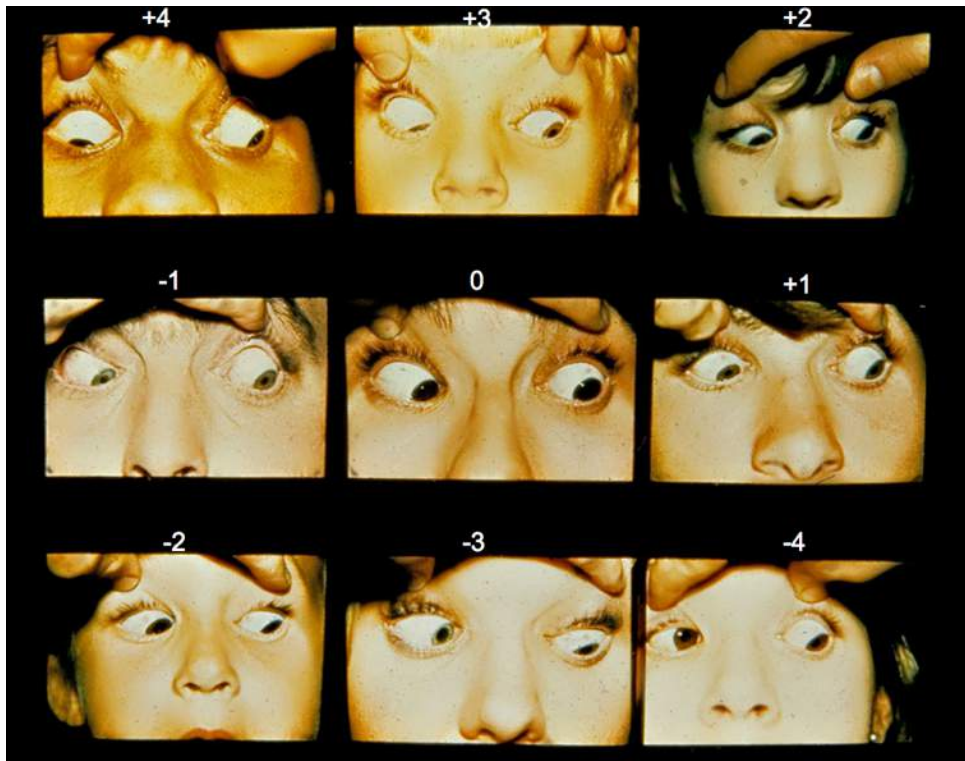
Grading Medial Rectus Versions (right eye)



Grading Lateral Rectus Versions (left eye)

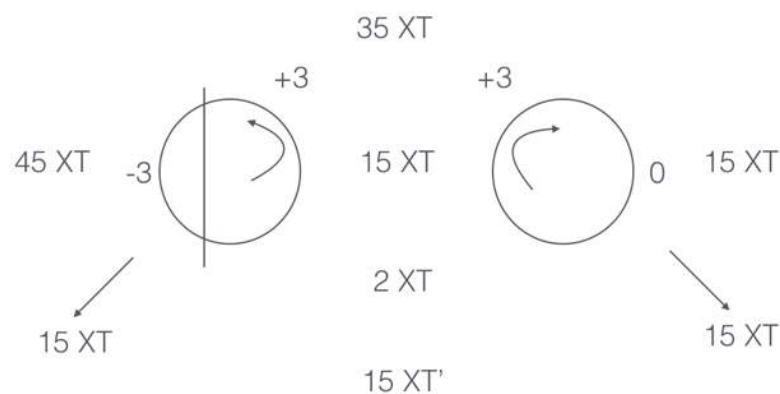


Grading Inferior Oblique Versions (right eye)



Grading Superior Oblique Versions (right eye)

- c. Differences on duction vs. version.
 - a. Versions > ductions, innervational problem
 - b. Ductions = versions, restrictive problem
 - d. End point or pathological nystagmus.
 - e. Changes in fissure size.
 - f. Lid changes.
8. Diagrammatic representation of versions
 -Versions and measurements should match.



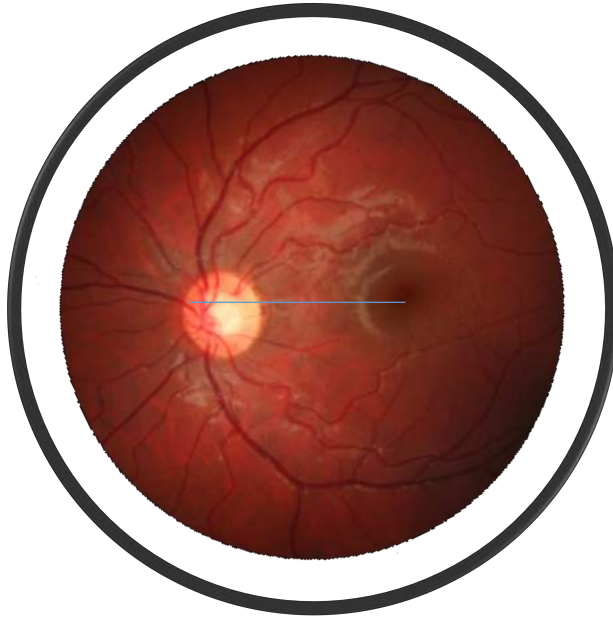
Example of recording measurements, versions and ductions on the same diagram

I. Fusional amplitude measurement

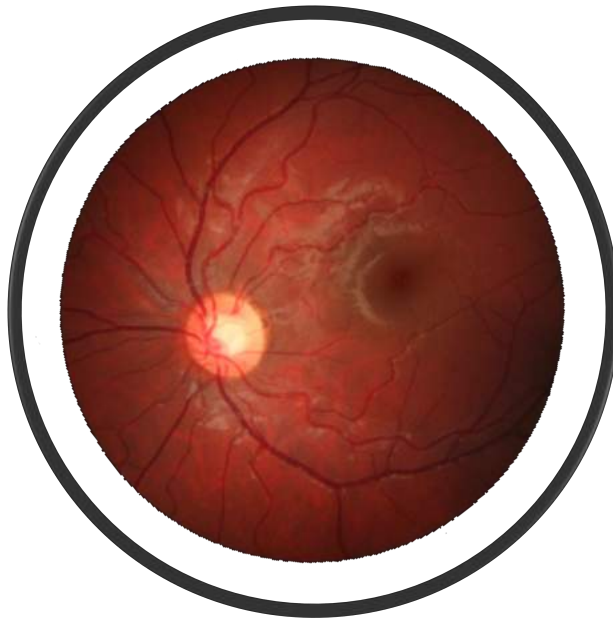
1. Convergence, divergence and vertical fusional amplitudes can be measured with a prism bar at both distance (20 feet) and near (13 inches). The bar is placed in front of one eye and is slowly moved from the smallest prism to prisms of increasing strength while the patient fixates a 20/40 target. The patient is instructed to report when the letter blurs and when he appreciates diplopia. At this point, the power of the prism is decreased until he again is able to fuse the image. The point where fusion broke, the one where it was recovered, as well as the blur point (which may not always be reported) are recorded. The blur point is the point at which the patient can no longer exert fusional convergence to overcome diplopia and tries to use accommodative convergence for this purpose. By accommodating in excess of the requirements for a given distance, the image will be blurred.
2. Fusional amplitudes cannot be measured on suppressing or diplopic patients. It is necessary to look for fusional movements while doing this test to ensure that the patient is not suppressing.

J. Torsion measurements

1. Double Maddox Rod
Orient both red and white lenses vertically, thereby creating two horizontal lines. Ask the patient to make both lines horizontal by turning the knobs at the sides of the frames.
2. Bagolini lenses
Less dissociative than #1. Orient both lenses at 90°. Lines will be created at 180°. Measure torsion as in #1.
3. Single Maddox Rod
Can only qualitatively assess torsion.
4. Maddox Double Prism
Can only qualitatively assess torsion. Consists of two 4^Δ prisms mounted base to base. The prism is placed before one eye. When looking at a single horizontal line, the patient will thus see three lines. The central line is seen by the eye without the prism while the Maddox double prism causes the other eye to see two lines - one above and one below the central line. The patient then judges whether or not the central line is parallel to the other lines.
5. Objective fundus viewing
 - a. With indirect ophthalmoscope.

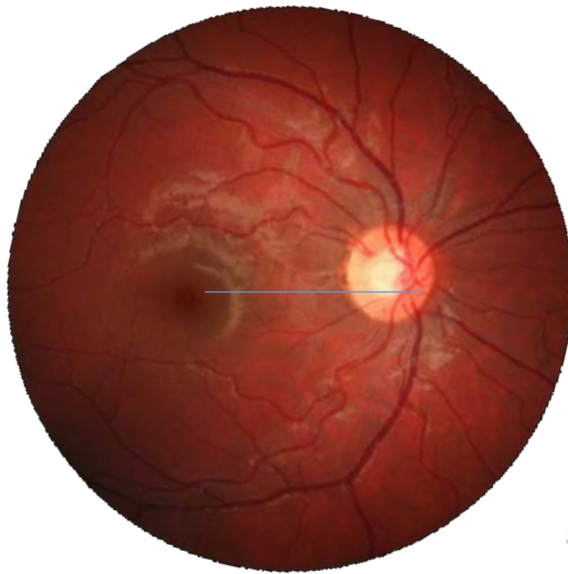


Normal Fundus Torsion of right eye seen with indirect Ophthalmoscope, the fovea falls within the upper 1/3 of the disc

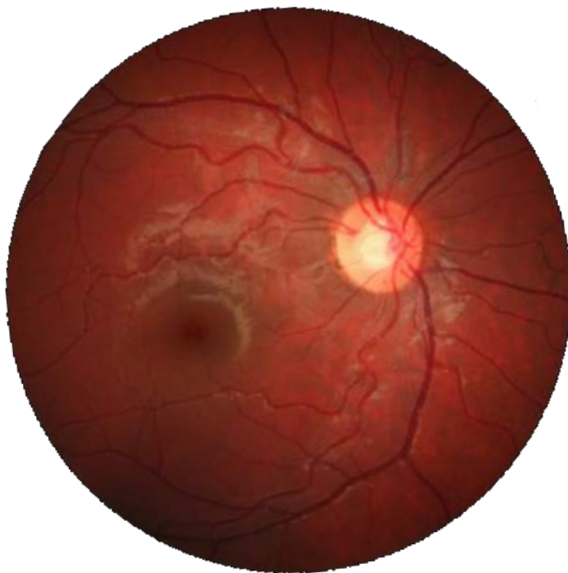


Excyclotorsion of right eye seen with the indirect ophthalmoscope, the fovea is above the disc

b. With fundus camera.



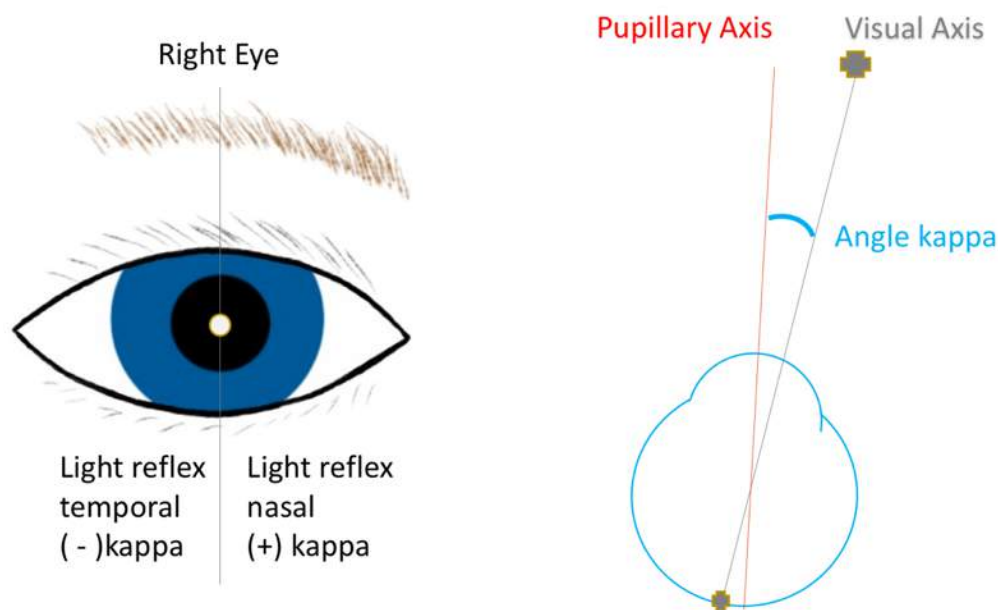
Normal fundus torsion of right eye seen on a fundus photograph, the macula falls within the lower 1/3 of the disc



Excyclotorsion of right eye seen in a fundus photograph, the macula is below the disc

K. Angles alpha, gamma, kappa and

1. Optic axis . The line that passes through the center of curvatures of all the refracting surfaces.
2. Pupillary axis. A line perpendicular to the cornea that passes through the center of the pupil. Usually coincides with the optical axis.
3. Visual axis. A line from the point of fixation to the fovea passing through the nodal points of the eye.
 - a. Angle alpha . Angle between optic axis and visual axis at the anterior nodal point.
 - b. Angle gamma. Angle between optic axis and visual axis at the center of rotation.
 - c. Angle kappa. Angle between pupillary axis and visual axis at the anterior nodal point. The angle that is clinically called the angle kappa is actually, the angle lambda.
 - d. Angle lambda. Angle between pupillary axis and visual axis at the center of the pupil.



Angle kappa

*Scott WE, Mash AJ. Kappa Angle Measures of Strabismic and Nonstrabismic Individuals. Arch Ophthalmol. 1973;89:18-20.

II. Examining the Diplopic Patient

A. A careful history should be taken to examine the patient's complaint.

1. Is diplopia monocular or binocular?
 - a. If covering one eye relieves the diplopia, make sure that is true for either eye.
 - b. Use a red filter and the distance fixation light to give the patient two different images at distance. Ask how many lights the patient sees and of what color. More than one of each color indicates that the patient has monocular diplopia.
2. Is diplopia worse at distance (when driving) or at near (when reading)?
3. Is diplopia constant, intermittent or variable?
4. Is diplopia horizontal, vertical, oblique or torsional?

B. Measure using the cover test providing the patient has adequate visual acuity for fixation.

1. Does this amount of prism alleviate diplopia?
2. If patient has a vertical and horizontal component to his/her deviation, will correcting one direction of the misalignment enable the patient to control the other component?

C. Use a red filter and the distance fixation light to measure the deviation. Can patient fuse?

1. ET - uncrossed (homonymous) diplopia.
2. XT - crossed (heteronymous) diplopia.
3. HT - object will be viewed as lower or below fixation target.
4. HypoT - object will be viewed as higher or above fixation target.
5. Torsion - object will be viewed as having a rotation opposite that measured.

D. Fields of binocular single vision

1. Diagnostic test for diplopic patients. Varies with gaze position and the eye chosen for fixation in incomitant patients.
2. Area of least deviation is characteristically diagonally opposite to the field of main action of the paretic muscle.
3. Can be performed on any perimeter used for monocular visual field testing. Unlike monocular field testing, the patient must follow the target with his eyes.
4. Must have:

- a. Normal monocular fields.
- b. Reasonable visual acuity without correction.
- c. No suppression.

Introduction to Strabismus

I. Incidence of Strabismus (Ages 1-74 years)

A. Heterotropias 3.7% (7.1 million)

1. Esotropia 1.2%
2. Exotropia 2.1%
3. Hypertropia 0.6%

B. Heterophorias 16.0% (30.7 million)
(From National Health Survey)

C. Prevalence of heterotropia in children

1. McNeil (1955) 2.7%
2. Frandsen (1960) 4.5%
3. Nordlow (1962) 3.86%
4. Graham (1974) 4.39%
5. Schutte, et al (1976) 4.3%

* Simons K, Reinecke R. Amblyopia screening and stereopsis. In: Symposium on Strabismus: Transactions of the New Orleans Academy of Ophthalmology. St. Louis; CV Mosby Co:1978, pp. 15-50.

* Graham PA. Epidemiology of strabismus. Brit.J. Ophthal. 1974;58:224-231

D. Incidence of heterotropia in children:

1. Mahoney (2007) 18.6 per 10,000 (0.19%) in a 10 year period

* Mahoney BG. Common forms of childhood strabismus in an incidence cohort. Am J Ophthalmol. 2007;144(3):465-7.

E. Prevalence of strabismus is 2-3% of the general population.

II. Frequency of Types of Strabismus (From private practice of Marshall Parks)

A. Esodeviations 60%

1. Accommodative 50%
 - a. Hypermetropic 40%
 - b. Pure High AC/A 10%

2. Non-accommodative 10%

B. Exotropia and verticals 40%

Esotropia

I. Definition

- A. Esophoria - a convergent deviation held latent by fusion.
- B. Intermittent Esotropia - a convergent deviation held latent at times but can be manifest.
- C. Esotropia - a manifest convergent deviation.

II. Etiology

A. Mechanical or anatomic

- 1. Secondary contractures
- 2. Adherence syndromes

B. Innervational

- 1. Excessive tonic convergence
- 2. High AC/A ratio

C. Refractive

D. Fusional deficiencies

E. Genetic (Multifactorial) 25-50% (Mash, Spivey)

F. Nystagmus

G. Impaired vision in one eye (Sensory, 4%)

- 1. If visual impairment is from birth, an esodeviation is most likely. If visual impairment is after age 3-4, eye will exodeviate.
- 2. If seeing eye is hyperopic, esodeviation is most likely; if closer to emmetropia, eye will exodeviate.

III. Classification

A. Congenital

1. Pseudostrabismus
2. Congenital (infantile) ET
3. Congenital non-comitant ET
 - a. Duane's syndrome
 - b. Moebius syndrome
 - c. Congenital VI nerve palsy

B. Acquired

1. Accommodative
 - a. Hypermetropic ET
2. High AC/A ratio
3. Combined High Hypermetropia, High AC/A ratio
4. Non-accommodative ET
 - a. High AC/A ratio very uncommon in this type of ET

D. Monofixation syndrome

IV. Work-up of a Patient with Esotropia

A. Sensory testing

B. Visual acuity - diagnosis of amblyopia

C. Versions

D. Measurements

1. Determine the Basic Deviation

E. Cycloplegic refraction

F. Fundus examination

V. Pseudoesotropia - Accounts for About 50% of All Suspected ET

A. Epicanthal folds, most common cause

1. Bilateral, usually asymmetrical
2. Three types of epicanthus
 - a. Supraciliaris
 - b. Palpebralis
 - c. Tarsalis

B. Large negative angle kappa, uncommon

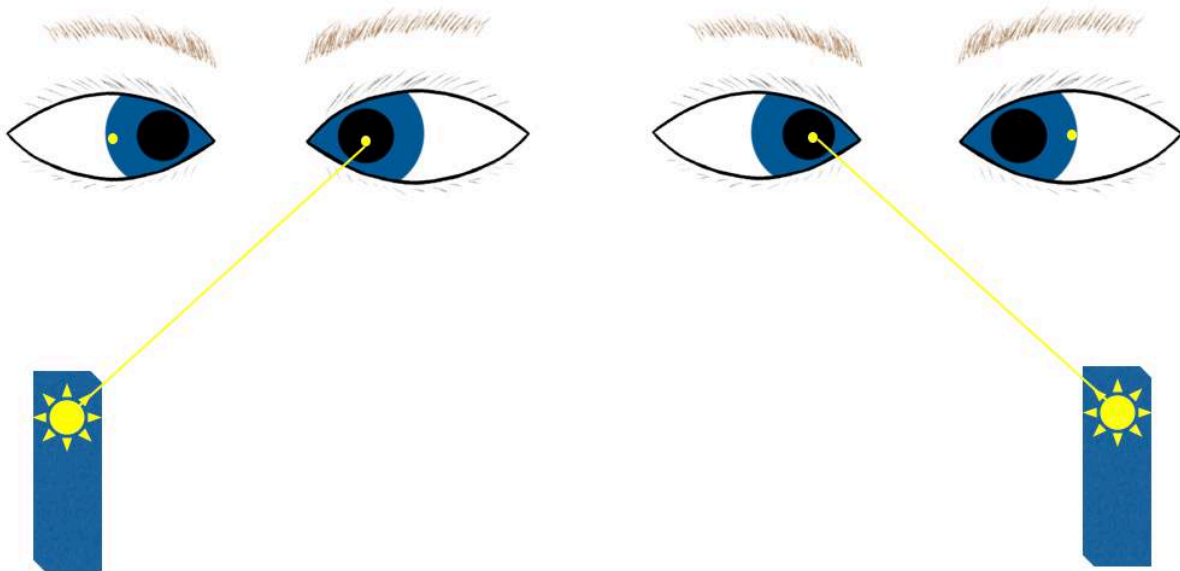
C. Abnormally small inter-pupillary distance

Congenital (Infantile) Esotropia

I. Characteristics

A. Characteristics

1. Onset before six months of age - generally not present at birth
2. Large angle of deviation
3. 40% prevalence of amblyopia
4. Crossfixation
 - a. The child fixates with the adducted eye for objects in the temporal visual field.
 - b. If the eyes switch fixation at midline, amblyopia is less likely present.



Crossfixation

5. Distance and near deviations are equal
6. Low refractive errors
7. Latent nystagmus 40-60%
8. DVD 60-80%
9. Inferior oblique over-action may occur.
10. V pattern

11. Peripheral fusion at best - not bifoveal fusers
12. 40-50% will develop an accommodative component
13. Strong family history

B. DVD - Dissociated Vertical Deviation: a spontaneous manifest vertical deviation

a.k.a.: Alternating Hyperphoria
 Double Hypertropia
 Occlusion Hyperphoria
 Alternating Sursumduction
 Dissociated Vertical Divergence

Occlusion Hyperphoria: a latent vertical deviation becoming manifest only when fusion is suspended.

1. Clinical characteristics
 - a. Usually bilateral - eye deviates up and out when dissociated.
 - b. Asymmetrical.
 - c. Variable amount and frequency of deviation.
 - d. If a fixation preference exists, a DVD may be present in the non-preferred eye with an occlusion hyper in the fixing eye.
 - e. Head tilt to the side of the DVD to control.
 - f. Usually associated with congenital esotropia or A pattern exotropia.
 - g. Rarely occurs without horizontal strabismus.
 - h. Rarely have bifoveal fusion.
2. Explanations of DVD
 - a. Bielschowsky (1930)
 - 1) Unilateral stimulation to upward gaze (variation, occlusion, and reduced illumination).
 - 2) Unequal stimulation of each retina. Abnormal excitability of the subcortical vertical divergence center (there is no evidence for these centers).
 - b. Posner (1944)
 Synthesis of the primitive monocular tonus regulator and the higher binocular innervation. Reflex mechanism - eyes tend toward divergence and elevation, i.e. Bell's Phenomenon.
 - c. Chavasse (1939) - One retina is less dominant - the eye moves up and out.
 - d. Verhoeff (1941) - Nucleus hypoplasia with overaction of obliques.
 - e. White & Brown (1939) - Due to paresis of SR, IR, or IO.
 - f. Guyton (1998)- exaggerated normal eye movement used to damp cyclovertical latent nystagmus
 - g. Brodsky (1999) - manifestation of primitive visuo-vestibular reflex

- h. Supranuclear origin evidence
 - 1) Coordinated nature of movement.
 - 2) Failure of direction of gaze to alter deviation.
 - 3) IO overactions may exist - evidence of palsy is lacking.

C. DVD vs. IOOA - differential diagnosis

1. DVD - features
 - a. Causes elevation in adduction.
 - b. Usually comitant, i.e. same in adduction, primary and abduction.
 - c. No corresponding hypotropia.
 - d. Variable hyper, at times appears small - moderate, other times large.
 - e. Generally not associated with a pattern.
 - f. Same amount of hyper in up-gaze and down-gaze.
 - g. Hyperdeviation may be associated with torsion and abduction.
2. Inferior Oblique Overaction - features
 - a. Causes elevation in adduction.
 - b. Incomitant - larger deviation in field of action of IO.
 - c. Not variable.
 - d. Commonly associated with a V pattern.
 - e. More hyper in up-gaze than in down-gaze.
 - f. Corresponding hypotropia.
3. Measurement
 - a. DVD - measure or estimate deviation in non-fixing eye.
 - b. IOOA - neutralize the hyper or hypotropia.
4. Treatment
 - a. IOOA - weaken IO.
 - b. IO weakening does little to correct DVD.
 - c. DVD - recess SR.
5. Treatment results
 - a. IO weakening corrects at most 15^A of vertical in primary, more in its field of action none in the opposite field.
 - b. SR recession cosmetically improves a DVD.
6. Previously used surgical procedures for the correction of DVD
 - a. 4mm superior rectus recession.
 - b. Inferior rectus resection.
 - c. Vertical R & R.
 - d. Faden or posterior fixation suture.
7. "Super Maximum" recession of SR is now the treatment of choice for DVD.
 - a. Technically easier than the Faden operation.

- b. Effective
- c. Does not cripple the muscle.
- d. Produces no lid changes.

8. Amount of SR recession for DVD

<10 ^Δ	5mm
<15 ^Δ	6mm
<20 ^Δ	7mm
>20 ^Δ	8mm
>25 ^Δ	9mm

*Braverman DE, Scott WE. Surgical treatment of dissociated vertical deviations. J Ped Ophthalmol Strab 1977; 14:337-342.

*Scott WE, Sutton VJ, Thalacker JA. Superior rectus recessions for dissociated vertical deviation. Ophthalmol 1982; 89(4):317-322.

*Guyton DL, et.al. Dissociated Vertical Deviation: An Exaggerated Normal Eye Movement Used To Damp Cyclovertical Latent Nystagmus. Tr. Am. Ophth. Soc. 1998;96:386-424.

*Brodsky MC. Dissociated Vertical Divergence. Arch Ophthalmol. 1999;117:1216-1222.

II. Management of Congenital Esotropia (developed over 16 years)

A. Preoperative evaluation

1. The goal in all strabismus surgery is to get the patient straight with the fewest number of operations.
2. Prior to surgery, the patient should be evaluated for amblyopia, refractive error, accommodative component, size of the deviation, and associated features.
 - a. Incidence of amblyopia is 40%, treated with full time occlusion of the better seeing eye.
 - b. Refractive status is evaluated after dilation with 1% Cyclogyl.
 - 1) $\geq +2.00$ D is given as a trial to observe its influence on the deviation.
 - 2) Refractions are repeated until two refractions agree within 0.5 D.
 - c. Stable prism cover test measurements are obtained at distance and at near.
 - 1) Two consecutive measurements that agree within 5^Δ.
 - 2) Deviation measured on an accommodative target at distance and near.
 - d. Complete examination includes investigation in the diagnostic positions of gaze.
 - 1) Rule out A or V pattern with oblique overaction or underaction.
 - 2) Differentiate inferior oblique overaction from DVD. Variable elevation in adduction caused by DVD.

B. Surgical plan

1. Selective surgery consists of operating on three or four horizontal muscles where esotropia exceeds 50^Δ.

Preoperative Deviation (Δ)	Bimedial Recession	Lateral Resection	Bilateral Resection
30 ^Δ	Leave 10mm from limbus		
35 ^Δ	Leave 10.5mm from limbus		
40 ^Δ	Leave 11mm from limbus		
45-50 ^Δ	Leave 11.5mm from limbus		
55 ^Δ	Leave 11mm from limbus	4-5mm	
60 ^Δ	Leave 11mm from limbus	6 mm	
65 ^Δ	Leave 11mm from limbus	7 mm	
70 ^Δ	Leave 11mm from limbus		4 mm
75 ^Δ	Leave 11mm from limbus		5 mm
80 ^Δ	Leave 11mm from limbus		6 mm
90-100 ^Δ	Leave 11mm from limbus		7 mm

2. Results of surgical alignment using the selective approach
 - a. Average pre-operative deviation in 48 patients who underwent three or four muscle procedures was 70^Δ.
 - b. Successful alignment was obtained in 65%.
 - c. Sensory fusion resulted in 40% of patients.

*Kraft SP, Scott WE. Surgery for congenital esotropia - an age comparison study. J Ped Ophthalmol Strab 1984; 21:57-68.

*Scott WE, Reese PD, Hirsh CR, et al. Surgery for large-angle congenital esotropia. Two versus three and four horizontal muscles. Arch Ophthalmol 1986; 104:374-377.

C. Variability of medial rectus insertion

1. Anatomically

Medial rectus insertion classically referred to as being 5.5mm from the limbus.

- a. Variability of medial rectus insertion has been reported by Helveston and Kushner. (See conventional surgery)
 - 1) Helveston measured distance between medial rectus insertion and limbus in 114 eyes; average 4.4mm (range 3-6mm).
 - 2) Kushner measured distance between medial rectus insertion and limbus in 80 eyes; average 4.3mm (range 3.5-5.5mm).
- b. Apt describes a change in the distance between medial rectus insertion and limbus with intraoperative manipulation.

*Apt L. An anatomical re-evaluation of rectus muscle insertions. Trans Amer Ophthalmol Soc 1980; 78:365-375.

2. Surgically
 - a. Measurement of insertion in 26 eyes of patients ranging in age between 10 and 30 months with preoperative deviation of 25^{Δ} - 70^{Δ} .
 - 1) A limbal conjunctival incision was used.
 - 2) Distance between the surgical limbus and anterior medial rectus insertion was measured.
 - 3) Average insertion 5.5mm from limbus (range 5-6mm).
 - b. Measurement of distance between the stump of the disinserted medial rectus and limbus.
 - 1) Following disinsertion, a measurement was taken of the distance between the limbus and the stump of the muscle.
 - 2) There was an average of 1.2mm movement of medial rectus muscle stump toward the limbus (range 0.5-2.0mm).
 - c. The muscle was re-attached to the globe an appropriate distance from the limbus using a curved ruler.
3. Following re-attachment, the distance between the stump of the medial rectus and the limbus was remeasured. The distance of the medial rectus muscle stump was an additional 0.5mm closer to the limbus.
4. Significant variability exists in the distance of the medial rectus muscle from the limbus secondary to intraoperative factors.
 - a. More accurate measurements of the amount of recession being performed are obtained by measuring from the limbus.
 - b. The location of the medial rectus muscle should be measured before disinsertion.

*Keech RV, Scott WE, Baker JD. The medial rectus muscle insertion site in infantile esotropia. *Amer J Ophthalmol* 1990; 109:79-84.

D. Conjunctival recessions to augment medial rectus recession

1. Helveston has reported that recession of the conjunctiva adds to the effect of medial rectus recession.
2. Guidelines used for recession of the conjunctiva
 - a. Grasp the eye with fixation forceps and abduct it.
 - b. With a small Stevens tenotomy muscle hook, palpate the conjunctiva with the eye abducted.
 - 1) If the conjunctiva is tight, it should be recessed back to the original insertion at the time of the surgery.
 - 2) If the conjunctiva is loose, recession will not augment the amount of deviation corrected with the medial rectus recession.
 - c. If there is tight conjunctiva, it is recommended that a limbal incision be used so the conjunctiva can be recessed.
 - d. Either a limbal or fornix incision may be used in the presence of loose conjunctiva.

III. Duane's Syndrome

A. Clinical characteristics

1. More common in females than males.
2. OS affected more often than OD.
3. 10%-30% incidence of anisometropic amblyopia.
4. Face turn toward the side of limited movement permits fusion.
5. 80% of cases are unilateral, when bilateral it is usually asymmetric.
6. Orthophoric, esotropic or exotropic in the primary position.
7. Angle of deviation always less than 30^{Δ} , usually less than 15^{Δ} .
8. Narrowing of the fissure and retraction of the globe of the involved eye on attempted adduction.
9. Abduction is limited in ortho and ET Duane's.
10. XT Duane's show limited adduction.
11. The amount of limitation depends on the amount of co-contraction of the medial rectus and lateral rectus.
12. Forced ductions are positive.
13. Abnormal firing of the lateral rectus is found with EMG testing.
14. Saccadic velocities on adduction are slowed secondary to the co-contraction.
15. Should be differentiated from a VI nerve palsy.

B. Alignment

- 36% orthophoric
- 25% exotropic
- 24% esotropic
- 15% bilateral

*Isenberg S, Urist MJ. Clinical observations in 101 consecutive patients with Duane's retraction syndrome. *Amer J Ophthalmol* 1977; 84:419-425.

C. Classification

1. Type I - poor abduction
normal adduction
2. Type II - ortho to exotropia
poor adduction
normal abduction
3. Type III - esotropia
poor abduction
poor adduction

D. Clinical-pathologic correlation

1. For example, in a left Duane's
 - a. The left abducens nucleus - no cell bodies from motor neurons except cell bodies compatible with internuclear neurons.
 - b. The left abducens nerve was absent.
 - c. The left lateral rectus muscle was partially innervated by branches from the inferior division of the IIIrd nerve.

*Miller NR, Kiel SM, Green WR, Clark AW. Unilateral Duane's retraction syndrome (Type 1). Arch Ophthalmol 1982; 100(9):1468-1472.

E. Surgical treatment of Duane's syndrome

1. Indications
 - a. Abnormal head position (AHP) - face turn toward limitation of movement.
 - b. Deviation in primary position.
 - c. Upshoot or downshoot on adduction.
2. Types
 - a. Type I - commonest of types; usually associated with AHP; esotropia in primary.
 - b. Type II - much less common; AHP.
 - c. Type III - more common than type II; usually not associated with AHP; upshoots or downshoots more common.
3. Surgical procedures
 - a. Single recession of MR if ET or LR if XT.
 - b. Recess MR - resect LR in involved eye.
 - c. Recess MR, recess LR in involved eye plus recess MR of normal eye with or without posterior fixation suture.
 - d. Transposition procedures temporally on involved eye.
 - e. Posterior fixation of LR.
 - f. Recession of LR with posterior fixation for upshoots and downshoots.

*Kraft SP. A surgical approach for Duane's syndrome. J Ped Ophthalmol Strab 1988; 25(3):119-130.

4. Surgical findings

- a. Forced duction positive to abduction in Type I.
- b. Forced duction positive to adduction in Type II.
- c. Tight medial rectus.
- d. Thin sclera under lateral rectus.

*Pressman SH, Scott WE. Surgical treatment of Duane's syndrome. Ophthalmol 1986; 93(1):29-38.

IV. Moebius syndrome (Congenital facial diplegia)

- A. No facial expression (VII nerve palsy)
- B. Esotropia (VI nerve palsy)
- C. Ptosis
- D. Deformity of external ear
- E. Atrophy of tongue
- F. Club foot, syndactyly, deficiency of pectoralis muscle.

Acquired Esotropia

I. VI Nerve Palsy

A. Clinical features in VI nerve palsy

1. Large angle esotropia in primary
2. Poor or absent abduction
3. Normal adduction
4. Absent firing or reduced firing of lateral rectus with EMG
5. Normal adduction on saccadic velocities

B. Differentiate from

1. Infantile esotropia with cross-fixation.
2. Duane's syndrome.

C. Congenital or acquired

1. Congenital
 - a. Birth trauma.
 - b. Hypoplasia of VIth nerve nucleus.
 - c. Anomaly of nerve fibers.
2. Acquired
 - a. Trauma to cranial floor.
 - b. Increased intracranial pressure.
 - c. Meningeal edema.
 - d. Inflammation in base of skull.
 - e. Displacement of brain stem.
 - f. Sensitivity to toxic substances.
 - g. Demyelinating disease.
 - h. Viral illness.
 - i. Vasculitis.

D. Treatment

1. Medical
 - a. Alternate occlusion while waiting for recovery
 - b. Fresnel prisms.
 - c. Botox- injected into antagonist muscle to help symptoms while waiting for recovery)

d. If recovery, usually in 3-6 mo.

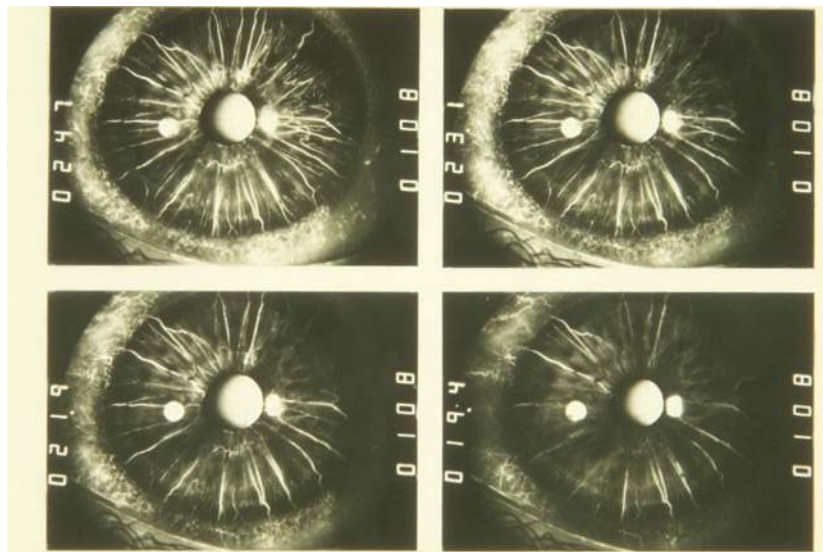
*Holmes JM et.al. Botulinum Toxin Verses Conservative Management in Acute Traumatic Sixth Nerve Palsy or Paresis. J AAPOS 2000;4(3):145-149.

2. Surgical

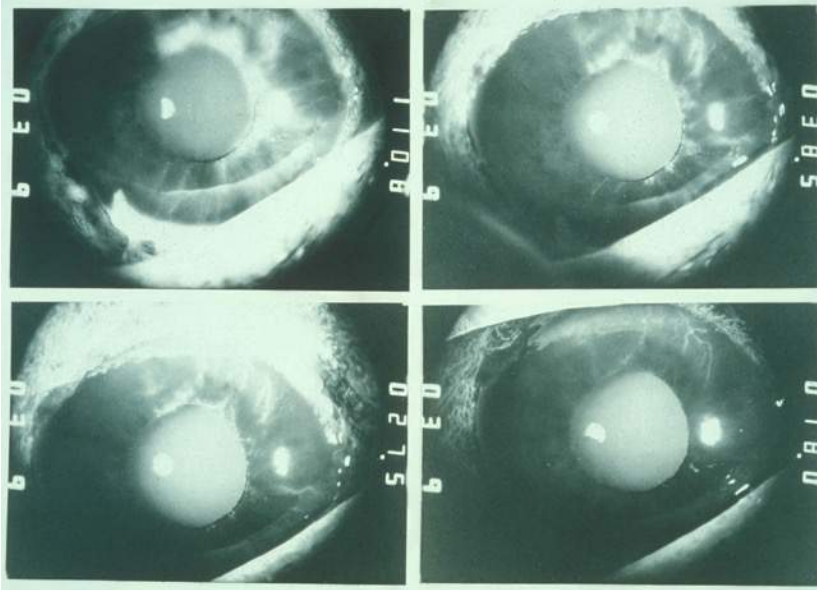
- a. If lateral rectus function is 40% of normal or better by saccadic velocity or force generation testing show poor function , R & R with adjustable suture.
- b. If lateral rectus function is absent or poor (less than 40% of normal),
 1. Jensen transposition procedure.
 2. Full tendon transposition and botox to medial rectus

E. Preoperative evaluation of VIth nerve palsy

1. Determine etiology
2. Exposure - cornea status
3. Diplopic field
4. Saccadic eye movement studies
5. Forced duction
6. Forced generations
7. Iris angiography if available to determine circulation changes to anterior segment



Normal Iris Angiography



Iris Angiography in Anterior Segment Ischemia (note segmental lack of iris vessel filling)

*Hayreh SS, Scott WE. Fluorescein Iris Angiography. I. Normal Pattern. Arch Ophthalmol 1978;96(8):1383-1389.

* Hayreh SS, Scott WE. Fluorescein Iris Angiography. II. Disturbances in Iris Circulation Following Strabismus Operation on the Various Recti. Arch Ophthalmol 1978;96(8):1390-1400.

F. Long term follow-up of Jensen procedures

1. Study population
 - a. 26 patients, 29 Jensen's procedures.
 - b. 9 bilateral, 17 unilateral.
 - c. Saccadic velocity less than 40% of normal (15 pts. less than 20% of normal).
2. Post-operative Results
 - a. 24/26 corrected to within 15^Δ of straight.
 - 1) Unilateral Jensen (n=20) 51^Δ improvement.
 - 2) Unilateral Jensen, bilateral surgery (n=3) 73^Δ improvement.
 - 3) Bilateral Jensen (n=3) 92^Δ improvement.
 - b. Improvement in saccadic velocity of 16.5%.
 - c. One case of anterior segment ischemia.
3. Summary
 - a. The Jensen procedure is stable over time.
 - b. Effectively provides fusion with acceptable head position.
 - c. Effective alignment with adjustable suture technique on the medial rectus.
 - d. Is safe with low incidence of anterior segment ischemia.
 - e. Comparison with other transposition procedures and R & R is still needed.

- * Cline RA, Scott WE. Long-term follow-up of Jensen procedures. J Ped Ophthalmol Strab 1988; 25(6):264-269.
- * Scott WE, Werner DB, Lennarson LW. Evaluation of Jensen procedures by saccades and diplopic fields. Arch Ophthalmol 1979; 97:1886-1889.

II. Accommodative Esotropia

A. History

1. Donders – 1864-
 - a. first to describe the association of hyperopia and esotropia
2. Duane's classification - 1924
 - a. Convergence excess - near deviation exceeds distance.
 - b. Divergence insufficiency - distance deviation exceeds near.
 - c. Combined - distance = near.
3. Costenbader – 1950
4. Parks - 1958
 - a. 50% of patients with ET have D-N disparity of 10^Δ or more.
 - b. High AC/A etiologic factor in acquired ET.

B. AC/A Ratio

1. Accommodative convergence - measured in meter angles or prism diopters
2. Accommodation - measured in diopters
3. Methods of determining the AC/A ratio
 - a. Heterophoric method -

$$AC/A = PD + \frac{\Delta n - \Delta 0}{D}$$

where Δn = measurement at near
 $\Delta 0$ = measurement at distance
 D = diopters of accommodation
 PD = pupillary distance
 - b. The heterophoric method assumes that convergence is wholly due to accommodation, it does not take into consideration the whole near response, i.e. tonic convergence, proximal convergence.
 - c. Gradient method - fixation distance is fixed; usually performed at near.
This is the Iowa preferred method.

$$AC/A = \frac{\text{change in deviation}}{\text{change in accommodation}}$$

The deviation is measured with and without a modifying lens. This lens changes the amount of accommodation used to create a clear image and thus changes the resultant deviation.

- d. Slope gradient method
Use series of lenses, i.e. +3.00, +1.00, -1.00, -3.00
Make slope of values.
- 4. Normal range of AC/A ratio
 - a. By gradient method - average is 3.7:1
 - b. Gradient method range is 0.9 - 9.8
 - c. By heterophoric method - average is 4.4:1
 - d. Heterophoric method range is 2.7 - 7.7 (0gle)
 - e. Gradient method
 - Low - 0-2.0
 - Normal - 2.5 - 5.0
 - High - >5.0
 - f. Factors that can influence the AC/A ratio
 - 1) Glasses/Bifocals
 - 2) Anticholinesterases
 - 3) Surgery
 - 4) Time
 - 5) Orthoptics

C. Types

- 1. Hypermetropic accommodative esotropia
 - a. One of the commonest types of accommodative ET
 - b. Distance and near measurements equal
 - c. Average age of onset 3.5 years
 - d. Average refractive error is +4.75 D
 - e. Amblyopia is common
 - f. Deterioration rate of 15% when previously controlled with glasses
- 2. Accommodative esotropia with a high AC/A ratio without significant hyperopia
 - a. Less than 1.50D of hypermetropia
 - b. Near deviation exceeds distance by 10^Δ
 - 1) Grade I - 10^Δ- 19^Δ greater at near.
 - 2) Grade II - 20^Δ- 29^Δ greater at near.
 - 3) Grade III - 30^Δ or more.
 - c. Amblyopia is common
 - d. Average age of onset 2 yr., 7 mo.
 - e. Rate of deterioration (Ludwig)
 - 1) Grade I - 25%

- 2) Grade II - 42.3%
- 3) Grade III - 51.6%
- f. Deterioration rate 30.3% overall
- g. Rate of deterioration increases with the severity of the AC/A and lower refractive error.
- h. Rate of deterioration is increased in patients with early onset of accommodative ET.

* Ludwig IH, Parks MM, Getson PR, et al. Rate of deterioration in accommodative esotropia correlated to the AC/A relationship. J Ped Ophthalmol Strab 1988;25(1):8-12.

- i. Deterioration rate - overall 13% (Dickey)
 - 1) AC/A ratio - no influence (only 12/93 > 9:1).
 - 2) Delay between onset and spectacle correction.
 - 3) Onset before 24 months of age.
 - 4) Decreasing hyperopia with > 5:1 AC/A.

* Dickey CF, Scott WE. The deterioration of accommodative esotropia: Frequency, characteristics, and predictive factors. J Ped Ophthalmol Strab 1988;25(4):172-175.

- 3. Combined hypermetropia - AC/A
 - a. Moderate hyperopia, +3.00 D
 - b. Distance-near disparity (N > D)
 - c. Average age of onset 3 years
 - d. Incidence of amblyopia is 30%
 - e. Commonest type of esotropia

D. Management of accommodative esotropia

- 1. Occlusion for amblyopia
- 2. Anti-accommodative therapy
 - a. Optical
 - 1) Single vision lenses: full hypermetropic correction.
 - 2) Bifocal lenses: to elicit fusion at near.
 - b. Medical - miotics. Optical correction is a more permanent solution.
- 3. Surgery for non-accommodative component

E. Indications for surgery in deteriorated accommodative esotropia

- 1. Unable to keep patient aligned with glasses alone
 - a. For fusion.
 - b. For alignment.
- 2. Once deterioration has occurred an attempt should be made to align patients and re-establish fusion pre-operatively with Fresnel prisms ("Prism Adaptation"). See Page 35

III. Mydriatics, Cycloplegics and Anticholinesterases

A. Mydriatics & cycloplegics

1. Cyclogyl (Cyclopentolate) - parasympatholytic
 - a. Peak cycloplegia in 40 minutes.
 - b. Compares favorably with atropine as a cycloplegic.
Robb and Petersen (1968) found less than 0.50 D more hypermetropia with atropine than 1% Cyclogyl.
 - c. Endpoint is two refractions that agree within 0.50 D.
 - d. Concentration/Dosage -
 $\frac{1}{2}\%$ - 2 drops at a 5 minute interval in children under age 1.
1% - 2 drops at a 5 minute interval in children over age 1.
2% rarely used.
Preferred method: 1 drop of 1% repeated in 5 minutes and 2.5% neosynephrine to enhance mydriasis in darkly pigmented individuals
 - e. Toxicity
 - 1) Flushing of the skin.
 - 2) Fever.
 - 3) Dryness of the mouth.
 - 4) Increased I.O.P.
 - 5) CNS - psychosis, behavior disturbances.
 - 6) Tachycardia.
2. Atropine - acts directly on smooth muscle
 - a. Onset - hours.
 - b. Duration - 10-14 days.
 - c. Concentration
1% - for darkly pigmented children.
 $\frac{1}{2}\%$ - for lightly pigmented children over age 4.
 $\frac{1}{4}\%$ - for lightly pigmented children under age 4.
 - d. Dosage - administered t.i.d. for 3 days prior to appointment; ointment should not be used on day of appointment.
 - e. Toxicity
 - 1) Fatal dose for children - 10 mg., 1 drop of 1% atropine = 0.5 mg.
 - 2) Systemic absorption is reduced with ointment.
 - 3) Reversed with physostigmine.
3. Mydracil (Tropicamide)
 - a. Peak effect in 20-25 minutes.
 - b. 30-40 minutes duration.
 - c. Produces less than 2.00D of residual accommodation.
 - d. Concentration/Dosage - 2 drops of 1% given 5 minutes apart.
 - e. Minimal toxicity.
4. Homatropine

- a. Onset - 10-30 minutes
- b. Peak cycloplegia in 3 hours
- c. Dosage - 6-8 drops given 10-15 minutes apart.
- d. Duration - 36 hours.
- e. Residual accommodation averages 1.00 D.

Agent	Instillation Procedure	Accommodation			
		Maximum Effect	Time of Effect	Able to Read	Normal
Cyclopentolate HCL (Cyclogyl)	1 drop x 2, 5 min. apart	25 min. after second drop	50 min.	3 hrs.	18 hrs.
Atropine Sulfate	1 drop, 3 times daily for 3 days	32 hours after first drop	8-24 hours	3-4 days	10-14 days
Tropicamide (Mydracyl)	1 drop x 2, 5 min. apart	20 min. after second drop	15 min.	45 min.	4 hours
Homatropine	6 drops x 2, 10-15 min. apart	40 min. after second drop	50 min	6 hours	36 hours

B. Anticholinesterases in accommodative esotropia- (rarely used in United States)

1. Mechanism
 - a. Inhibit or inactivate acetylcholinesterase.
 - b. Allows acetylcholine to accumulate at cholinergic receptor sites-acts as if cholinergic neurons are being continually stimulated.
2. If an anticholinesterase has to be given more than every other day to yield a good result, its use should be reconsidered.
3. Dosages of formerly used miotics
 - a. Diisopropyl Fluorophosphate (D.F.P.) - supplied as .025% ointment.
 - 1) Diagnostic dose 1/4 inch ointment every night for 2 weeks.
 - 2) Therapeutic dose - every night for two weeks; decrease to every other night for two weeks then once a week for 2 months.
 - b. Phospholine Iodide (P.I.) - stable longer than D.F.P.
 - 1) Diagnostic dose-0.125% every night for 2-3 weeks.
 - 2) Therapeutic dose-0.125% every other night.
 - 0.06% every night.
 - 0.03% every night.
 - c. Humorsol - not frequently used in strabismus.

4. Systemic effects of anticholinesterase agents.
 - a. Salivation.
 - b. Sweating.
 - c. Urinary incontinence.
 - d. Diarrhea.
 - e. Muscle weakness.
 - f. Respiratory difficulties (anticholinesterases are contraindicated in asthmatics).
 - g. Cardiac irregularities.
5. Ocular effects - miosis, ciliary spasm.
 - a. Pain - brow ache, retro-ocular ache.
 - b. Myopia.
 - c. Retinal detachment - never reported in children.
 - d. Lens changes - begin as mossy anterior subcapsular opacities; Later - nuclear sclerosis. When drug is discontinued, lens opacities seem to reverse but often appear later.
 - 1) Michon, Kinoshita (1967) -lens epithelium contains cholinesterases
 - 2) Leopold (1975) -anticholinesterases increase serum amino acids which may cause defects in lens metabolism.
 - e. Iris cysts-prevented by compounding with neosynephrine.
 - f. Ocular effects decrease after using the drug for two weeks.
 - g. Existing uveitis is a contraindication to the use of anticholinesterases.

IV. Non-Accommodative Acquired Esotropia

A. Clinical features

1. Moderate angle of deviation
2. Equal vision
3. Little or no hypermetropia
4. Normal AC/A Ratio
5. May show suppression or ARC

B. Patients falling into this classification are probably broken-down monofixators.

C. These patients deserve a complete neurological evaluation and consideration of CNS imaging

D. Treatment

1. Prism adaptation

2. Surgery for full amount of deviation

V. Prism Adaptation

A. Background

1. Indications for surgery in deteriorated accommodative esotropia: unable to align or keep patient aligned with glasses alone.
 - a. For fusion.
 - b. For alignment.
2. Indications for prism adaptation.
 - a. Deteriorated accommodative esotropia, deviation initially controlled by medical treatment.
 - b. Partially accommodative esotropia: glasses correct only part of the deviation.
 - c. Non-accommodative acquired esotropia.
3. Prism adaptation: preoperative test to predict fusional capability and success of surgery in acquired esotropia when surgery is indicated.

B. Technique of prism adaptation (PA)

1. Esotropia is neutralized distance and near with Fresnel press-on prisms; enough prism should be mounted to render patient orthotropic to slightly exotropic.
2. Patients are followed weekly and prisms adjusted appropriately until they can be labeled “responder” or “non-responder”.
 - a. Responder
 - 1) Deviation stabilized at 8^{Δ} esotropia or less with peripheral fusion on Worth 4-Dot while wearing prisms.
 - 2) In these patients surgery is done for amount of esotropia present after prism adaptation.
 - b. Non-responder
 - 1) With Fresnel prisms deviation builds to greater than 60^{Δ} without fusion.
 - 2) Angle of deviation does not build with prisms but fusion is not obtained; with prisms patient is exotropic at distance and esotropic at near without fusion.
 - c. In these patients, surgery is done for original angle of deviation.

C. Prism response

1. Deviation on simultaneous prism and cover test (SPCT) 0 to 8^{Δ} at distance and near plus fusion of Worth 4-Dot at $\frac{1}{3}$ m, or
2. Diplopia on Worth 4-Dot at near but also had 2/9 circles and 2/3 animals on Titmus stereo.

D. Prism non-response

1. Exotropia on SPCT at distance and near with suppression to Worth 4-Dot, or
2. Esotropia 0 to 8^Δ distance and near with no fusion; held here for 30 days
3. Deviation built to >60^Δ

E. Mechanism of prism adaptation

1. Uncovers latent esotropia
2. Measures fusion potential

F. Randomized clinical trial compared overall effectiveness of PA as preoperative test

G. Surgery

1. Target angle - amount of surgery done for esotropia in distance measured by ACPT at time of randomization
2. Used for
 - a. Control group.
 - b. Non-responders.
 - c. Prism responders randomized (level 2) to entry angle.
3. Prism adapted surgery done for total amount of prism at time of response to prisms.
4. Standardized to amount and technique.
5. Surgical table:

Target Angle Prism Diopters	Medial Rectus Recess Both Eyes	Lateral Rectus Resect One Eye
12 ^Δ - 15 ^Δ	3.0 mm	--
16 ^Δ - 20 ^Δ	3.5 mm	--
21 ^Δ - 25 ^Δ	4.0 mm	--
26 ^Δ - 30 ^Δ	4.5 mm	--
31 ^Δ - 35 ^Δ	5.0 mm	--
36 ^Δ - 40 ^Δ	5.5 mm	--
41 ^Δ - 45 ^Δ	6.0 mm	--
46 ^Δ - 50 ^Δ	5.0 mm	5.0 mm
51 ^Δ - 55 ^Δ	5.0 mm	6.0 mm
56 ^Δ - 60 ^Δ	5.0 mm	7.0 mm

6. Surgical Table: recess/resect procedures

<u>Target Angle Prism Diopters</u>	<u>Medial Rectus Recess</u>	<u>Lateral Rectus Resect</u>
15 ^Δ	3.0 mm	4.0 mm
20 ^Δ	3.5 mm	5.0mm
25 ^Δ	4.0 mm	6.0 mm
30 ^Δ	4.5 mm	7.0 mm
35-40 ^Δ	5.0 mm	8.0 mm
> 40 ^Δ see First Surgical Table above		

H. Success defined as 0 - 8^Δ by SPCT

I. Primary goal

1. Overall PA success rate, 83%
2. Success of controls, 72%
3. Statistically significant Z-statistic of 2.02 (p = 0.04)

J. Postoperative alignment

1. Total seven overcorrections (>8^Δ exotropia), five in controls (non-PA, surgery for entry angle).
2. Undercorrections (>8^Δ esotropia)
 - a. 25% controls.
 - b. 21% PA response/ES (surgery for entry angle).
 - c. 10% PA response/PS (surgery for amount of prisms).
3. Orthotropic
 - a. 49% PA response/PS.
 - b. 24% PA non-response/ES.

K. Once PA response, surgery for entry angle or surgery for prism adapted angle.

L. Summary

1. Prism adaptation had overall effect of producing better postoperative alignment than controls (p = 0.04).
2. Operating for built angle had lower rate of undercorrections (10%) without increasing overcorrections (<1%).

3. PA identifies group of patients that can safely receive larger amounts of surgery.
 4. Non-responders had lowest rate of postoperative fusion.
 5. 55% of prism responders built their angle little with prisms.
 6. Recommend that PA be done for patients with acquired esotropia to determine target angle for surgery.
- * Prism Adaptation Study Group. Efficacy of prism adaptation in the surgical management of acquired esotropia. Arch Ophthalmol 1990;108(9):1248-1256.
- * Repka MX et. al. The One-year Surgical Outcome after Prism Adaptation for the Management of Acquired Esotropia. Ophthalmology 1996;103:922-928.

VI. PA for Near Angle

A. When near deviation is greater than distance deviation

B. Preliminary results

1. Group I (n = 10), no fusion response to prisms or exotropia in distance with prisms
 2. Group II (n = 17) stabilized with prisms for near angle and fused
 3. Group III (n = 4) increased esotropia 10^A or more in response to base-out prism; no exotropia in distance
 4. Summary
 - a. 21 of 31 fused with prisms pre-op when near deviation was offset without any exotropia at distance.
 - b. 20 of 21 maintained fusion at last post-op exam.
 - c. None required bifocals post-op for fusion.
- * Kutschke PJ, Scott WE, Stewart SA. Prism adaptation for esotropia with a distance-near disparity. J Pediatr Ophthalmol Strabismus 1992;29(1):12-15.

VII. Esotropia Secondary to a Monocular Visual Defect

A. Characteristics

1. Secondary to structural abnormality
2. High anisometropia

3. Stimulus deprivation amblyopia (amblyopia is often dense and irreversible)

4. Poor fusion potential

B. Deviation is usually variable and tends to exodeviate over time.

C. Management of sensory ET

1. Maximum development of visual acuity.

2. Vigorous trial of occlusion therapy.

3. Full cycloplegic refraction.

4. Surgery.

VIII. Factors Complicating the Course and Treatment of Esotropia.

A. Amblyopia

B. Organic monocular visual deficits

C. Vertical deviations

D. Incomitance

E. Oblique dysfunction

F. DVD

G. A-V patterns

H. CNS abnormalities

I. Adhesive syndrome (A.K.A. Fat Adherence)

Monofixation Syndrome

I. Characteristics

- A. Deviation of 0^{Δ} - 8^{Δ} . An "orthotropic" monofixator has no shift on prism cover test but responds like a tropic patient to sensory testing.
- B. Central suppression of the deviated eye
- C. Good fusional vergence amplitudes
- D. Synonyms for monofixation syndrome
 - 1. Esophoria with retinal slip
 - 2. Esophoria with fixation disparity
 - 3. Fixational disparity
 - 4. Subnormal binocular vision
 - 5. Small angle esotropia
 - 6. Convergent fixation disparity

II. Features

- A. Constant features
 - 1. Absolute facultative scotoma in the monofixating eye
 - 2. Peripheral fusion
 - a. Fusion of Worth 4-Dot lights at near.
 - b. 3000 to 60 seconds of stereoacuity.
- B. Variable features
 - 1. Strabismus history
 - 2. Anisometropia
 - 3. Unilateral macular lesion
 - 4. Amblyopia

5. Eccentric fixation
6. Orthophoria
7. Phoria
8. Small tropia
9. Alternate cover exceeds cover-uncover

III. Etiology

- A. Treated strabismus
- B. Anisometropia
- C. Unilateral macular lesion
- D. Inherent inability to fuse similar images on each macula (primary monofixation)

IV. Tests for Monocular Scotoma During Binocular Viewing (Sensitivity)

- A. Worth 4-Dot Distance (100%)
- B. Stereopsis, <40 seconds (100%)
- C. Binocular perimetry (99%)
- D. Bagolini lenses (93%)
- E. 4^Δ Base Out (72%)

V. Specific Characteristics

- A. Amblyopia
 1. Incidence of amblyopia varies with the etiology of monofixation
 - a. Congenital esotropes - 34% incidence of amblyopia.
 - b. Acquired esotropes - 67%.
 - c. Primary monofixators - 73%.
 - d. Strabismus and anisometropia - 88%.
 - e. Anisometropes - 100%.

2. Most monofixators have amblyopia before treatment.

B. Ocular alignment in monofixation

1. 63% have a shift on cover-uncover test
 - a. 1^{Δ} - 8^{Δ} horizontally.
 - b. 2^{Δ} - 3^{Δ} vertically.
2. 37% have no shift on cover-uncover test
3. Deviation varied with etiology
4. Lack of shift on cover-uncover provides four possibilities
 - a. Monofixational orthophoria.
 - b. Monofixational phoria.
 - c. Bifixational orthophoria.
 - d. Bifixational phoria.
5. Orthophoria does not automatically indicate bifixation
6. Alternate cover often exceeds cover-uncover

C. Fusional divergence amplitudes in monofixation

1. Equal to those in bifixators. Approximately 7^{Δ} at distance and 10^{Δ} - 12^{Δ} at near.
2. Strabismic patients (exceeding 8^{Δ} deviation) have limited or non-existent amplitudes.

VI. Treatment

A. Motor

1. Surgery - rare.
2. Prisms - rare.

B. Sensory

1. Amblyopia therapy.
2. Orthoptics - not appropriate.
3. Follow every 6 months until age 8.

VII. Prognosis

A. Stability of monofixation syndrome (MS)

1. Average follow-up 17.5 years.
2. 74% remained aligned with MS.
3. 45% remained aligned without MS fusion.

B. Potential for bifixation – poor

* Parks MM. The monofixation syndrome. Trans Amer Ophthalmol Soc 1969;67:609-657.

C. Stability of alignment in monofixation- Probably the best desired result in the treatment of Congenital Esotropia.

1. Comparison of 38 patients with monofixation to 42 patients without monofixation. All well aligned ($\pm 8^\Delta$) following surgery for congenital esotropia.
2. No difference between those with monofixation and those without in terms of
 - a. Post-operative follow-up time.
 - b. Pre-operative esotropia.
 - c. Alignment immediately post-operatively.
3. Monofixators were aligned at a significantly earlier age (mean 2.6 yrs vs. 3.9 yrs).
4. Stability of alignment is significantly better in monofixators (estimated mean time at loss of stability 32.2 yrs vs. 9.8 yrs).

* Arthur BW, Scott WE. Long-term stability of alignment in the monofixation syndrome. J Ped Ophthalmol Strab 1989;26(5):224-231.

Exotropia

I. Classification

A. Exophoria - X

Tendency toward divergence of the visual axes held latent by fusion.

B. Intermittent Exotropia - X(T)

Tendency toward divergence of the visual axes not always controlled by fusion.

C. Constant Exotropia - XT

Constant divergence of the visual axes.

D. Duane's classification of exotropia

1. Divergence excess - larger deviation at distance than near.
2. Convergence insufficiency - larger deviation at near than distance.
3. Basic - distance and near deviations equal.
4. Pseudodivergence excess - apparent divergence excess, but when fusional convergence is eliminated with 45 minute occlusion and/or accommodative convergence eliminated with +3.00 lenses, near deviation equals distance.
5. Duane's classifications are used to plan the type of surgical correction.

II. Characteristics

A. Monocular

B. Alternating

C. Natural - 2/3rds of cases spontaneously occur as exotropia

D. Secondary - following surgery for esotropia

E. Consecutive - spontaneous XT usually following esotropia associated with high hypermetropia.

III. Etiology

A. Anatomical - position of rest of the eyes; exodeviation may worsen with age.

B. Heredity- increased risk in 1st degree relatives

C. Sensory abnormalities

1. Rare to find sensory abnormalities in exophoria.
2. Intermittent XT's usually only have a mild difference in visual acuity.
3. Temporal Suppression or ARC, once established tend to make the exotropia occur more easily. Since these sensorial adaptations develop over time, many XT's don't show up until the age of 6-8 years.

D. Innervational factors

1. Nerve palsy
2. Duane's syndrome
3. Decreasing AC/A ratio with age
4. Relaxation of the vergence mechanism

IV. Exophoria

A. A small exophoria is considered physiologically normal.

- a. Up to 4^{Δ} at distance and up to 6^{Δ} at near.
- b. No temporal suppression

B. Convergence insufficiency type exophoria - 2 types

1. "Eye Strain" associated with near work
 - a. X at near greater than distance.
 - b. Poor convergence amplitudes;
Normal amplitudes: near = 30^{Δ} - 40^{Δ} ; distance = 20^{Δ} - 30^{Δ} .
 - c. Reduced near point of convergence: normal range 4cm - 10cm.
 - d. Normal near point of accommodation.
 - e. This type of convergence insufficiency is treated with orthoptic exercises to teach convergence and build fusional amplitudes.
2. Hypo-accommodative convergence insufficiency
 - a. Premature loss of ability to accommodate - may be related to effort.
 - b. Treat with plus lenses for near work before attempting orthoptic therapy.

V. Intermittent exotropia

A. Onset usually between 0-10 years, peak incidence 5.5 years.

B. Progression

1. 20% progress to constant deviations.
2. A patterns tend to progress.
3. V patterns tend to be stable.
4. Convergence insufficiency type deviations tend to progress.
5. Divergence excess (distance deviation greater than near) type deviations tend to be stable.
6. X(T)'s that worsen usually do so after age 4-5 years.

C. Symptoms

1. Eye alignment deformity
2. Frequently one eye closes in bright sunlight.
3. Worse with fatigue or illness
4. Diplopia - usually only after some form of orthoptic treatment; i.e. anti-suppression
5. Family history often positive

D. Signs

1. Anisometropia
2. Minimal difference in visual acuity ($\frac{1}{2}$ to 1 line)
3. Patterns in approximately 30% of cases
4. Abnormal distance-near relationship
5. Usually strong preference for one eye
6. Temporal suppression
7. Differentiate constant and intermittent by controlling accommodation.
 - a. A constant exotropia can appear to be intermittent if they are using accommodation to control the alignment.

E. Follow-up (small to moderate angles)

1. Measure amount of deviation at distance and near on accommodative target. The change in alignment with accommodation effort can make a constant exotropia look intermittent.
2. Record frequency at home and on clinical exam of deviation (progression toward constant XT).
- 3 Lateral incomitance
4. Pattern
5. Approximately 40% will develop a vertical deviation.

F. Treatment

1. Treat amblyopia if more than 2 lines difference between eyes
2. Minus lenses to stimulate accommodation to control deviation. Most practical in glasses wearers
3. Prisms if angle is small enough
3. Orthoptics in an attempt to break temporal suppression
5. Patching (see occlusion below)
6. Surgery - these patients do well with bilateral lateral rectus recessions.

VI. Exotropia

A. Congenital exotropia

1. Onset at birth - congenital XT is uncommon.
2. Alternating - if a congenital XT is not alternating, further evaluation of etiology is indicated.
3. Usually large deviation
4. Requires large amounts of surgery for alignment
5. Poor fusional potential

B. Tight lateral rectus syndrome

1. Characteristics
 - a. Limitation of adduction.
 - b. Apparent overaction of all oblique muscles.
 - c. X pattern.
2. X patterns usually occur in longstanding exodeviations.
3. Large angle of deviation
4. More common in monocular exotropia
5. Recessing both lateral rectus muscles will treat the apparent oblique dysfunction.

C. Right angle exotropia

1. In primary position, the cornea of the deviated eye touches the lateral margin of the orbit.
2. Right angle limitation of adduction
3. Right angle limitation in vertical movements
4. Surgery - Urist (1964) was the first to recommend 4 muscle surgery for this type of exotropia

D. Exotropia associated with amblyopia

1. Onset usually in adults.
2. Surgery - large recess/resects (8-12 mm).

E. Vertical deviations associated with exotropia are common

1. If a vertical is present, there is usually a tendency to undercorrect.
2. If associated with amblyopia, determine if it causes an eye alignment deformity.
3. Determine with prisms, whether vertical is significant; if a patient is horizontally aligned with prisms he may be able to control the vertical.

Management of Exotropia

I. Non-Surgical Treatment - for XT of 15^{Δ} - 20^{Δ} or less.

A. Dominant eye occlusion

1. Works best in patients between 2-4 years of age
2. Comitant deviations less than 30^{Δ}
3. Patch dominant eye 4 hours to $\frac{1}{2}$ waking day for 3 weeks
4. Should improve control of deviation

B. Prisms

1. Break down suppression
2. Hardesty (1972) advocates use of base-in prism following surgical undercorrection, enough prism is used to keep patient from suppressing.
3. Prisms are usually worn 6 months - 1 year
4. 50% of patients wearing prisms do not need further surgery

*Hardesty HH. Prisms in the management of intermittent exotropia. Am Orthopt J. 1972;22:22-30

C. Minus lenses

1. Stimulates accommodative convergence - only practical in young children
2. Prevents suppression

D. Orthoptic therapy

1. Breakdown suppression
2. Improve NPC
3. Improve fusional amplitudes
4. Most effective on angles of 20^{Δ} or less
5. Not indicated in surgical candidates

E. Botulinum toxin

1. Injected into lateral rectus muscles
2. Produces temporary esotropia
3. Suppression breaks down
4. Control of deviation improves

II. Surgical Treatment

A. Pre-operative evaluation

1. Determine basic deviation
2. Determine type of control, i.e. constant distance and near; XT distance, X(T) near, etc.
3. Look for patterns
4. Lateral gaze measurements - measure in mid-right and mid-left gaze; take note of extreme right and left gaze deviation.
5. Measure at near with +3.00 D lenses to eliminate accommodative convergence.
6. Measure after 45 minute occlusion.
7. Assess versions - real vs. pseudo oblique overactions, limitation of adduction, tight LR's

B. Type of surgery depends on

1. Age of patient
2. Type of deviation
3. Visual acuity
4. Pattern
5. Measurements
6. Versions

C. Surgical choices for exodeviations

1. Recession/resection

- a. Older patients (>8 years).
 - b. Deviation equal distance and near.
 - c. Constant XT distance and near.
2. Bilateral lateral rectus recessions
 - a. Patients less than 8 years of age.
 - b. Distance deviation greater than near.
 - c. Intermittent deviation.
 3. Bimedial rectus resections
 - a. Rarely done
 - b. Convergence insufficiency type exodeviations.

D. Surgical goals

1. Temporarily overcorrect to break down suppression, leave patients 4^{Δ} - 14^{Δ} ET
2. Cure = phoria with no evidence of suppression

E. Dosage of surgery for exotropia

<u>Preoperative Deviation ($^{\Delta}$)</u>	<u>Bilateral Recessions</u>	<u>Recess/Resect</u>
15	4 mm	4/3 mm
20	5 mm	5/4 mm
25	6 mm	6/5 mm
30	7 mm	7/6 mm
35	8 mm	8/6 mm

<u>Preoperative Deviation ($^{\Delta}$)</u>	<u>Bilateral LR Recessions</u>	<u>Resect One Medial Rectus</u>
40	7 mm	4 mm
45	7 mm	5 mm
50	7 mm	6 mm
		<u>Resect Both Medial Recti</u>
55	7 mm	4 mm
60	7 mm	5 mm
65	7 mm	6 mm
70	7 mm	7 mm
80	8 mm	7 mm

Use adjustable suture whenever possible on 1 LR up to 60^{Δ} , on both LR on deviations $>60^{\Delta}$.

If operating on a patient with amblyopia, do recess-resect on amblyopic eye.

<u>Preoperative Deviation (Δ)</u>	<u>Recess/Resect</u>
40	8/7 mm
50	9/7 mm
60	10/8 mm
70	10/9 mm
80	10/10 mm

F. Management of surgical undercorrections which are more common than overcorrections

1. Prisms.
2. Minus lenses.
3. Orthoptics.
4. A second surgery may be necessary.

* Scott WE, Keech RV, Mash AJ: The postoperative results and stability of exodeviations. Arch Ophthalmol 1981;99:1814-1818.

Patterns Associated with Strabismus

I. Types of Patterns

- A. "V" Patterns - increased esodeviation or decreased exodeviation in down-gaze. 15^Δ difference in up-gaze vs. down-gaze
- B. "A" Patterns - increased esodeviation or decreased exodeviation in up-gaze. 10^Δ difference in up-gaze vs. down-gaze
- C. "X" Patterns - more exodeviation in up-gaze and down-gaze than in primary position
- D. "Y" patterns - marked divergence in up-gaze, less in primary and down-gaze
- E. "V" Patterns are 5 times more common than "A" patterns

II. Etiology

- A. Anatomic theory - (Urrets-Zavalía)
Local variations in muscle attachment and orbital configuration.
- B. Innervational theory - theoretical CNS center alters muscle tone.
- C. Horizontal muscle tone theory - (Urist)
MR adductors, especially in down-gaze; LR abductors especially in up-gaze. Patterns are caused by overactions or underactions of these muscles.
- D. Theoretical surgical considerations based on the horizontal muscle theory
 - 1. "V" eso – bilateral medial rectus recessions
 - 2. "A" eso - bilateral medial rectus resections
 - 3. "V" exo – bilateral lateral rectus recessions
 - 4. "A" exo – bilateral medial rectus resections
- E. Vertical rectus theory - (Brown)
The SR and IR are both adductors and their underaction or overaction could cause a pattern.
- F. Theoretical surgical considerations based on the vertical rectus theory - (Miller)
 - 1. "V" eso - transplant IR 7 mm temporally

2. "A" eso - transplant SR 7 mm temporally
3. "V" exo - transplant SR 7 mm nasally
4. "A" exo - transplant IR 7 mm nasally

G. Horizontal-vertical theory - (Tamler) compromise theory.

1. Pure syndrome
 - a. No vertical muscle imbalances.
 - b. Rx - only horizontal muscle surgery with change in height of insertion.
2. Impure syndrome
 - a. Associated vertical muscle anomalies.
 - b. Rx - combined horizontal and vertical muscle surgery.

H. Oblique muscle theory - (Jampolsky)

Patterns are due to the abducting effect of the IO's in up-gaze and the SO's in down-gaze.
This is the most popular and widely held theory.

I. Theoretical surgical considerations based on the oblique muscle theory.

1. "V" eso - IO weakening
2. "A" eso - SO weakening
3. "V" exo - IO weakening
4. "A" exo - SO weakening

J. Objections to obliques being the sole cause of patterns (Burian, Cooper, Costenbader)

1. Surgery on the horizontal recti may influence or even cause a pattern.
2. Patterns may occur in the absence of appropriate oblique overactions.
3. Obvious over and underactions do not always result in a pattern.
4. Appropriate oblique surgery may not cure a pattern.

III. Principles of Treatment

- A. Divide patients into those with oblique dysfunction and those without. Majority will have oblique dysfunction.
- B. Select surgery to reduce horizontal deviation in primary position and minimize A-V incomitancy.
- C. Horizontal muscle shift works well when there is no oblique dysfunction. It is not a substitute for oblique surgery.
- D. Weakening IO's or tuck of SO's each correct 15^{Δ} - 25^{Δ} of a "V" pattern.
- E. Bilateral superior oblique tenotomies correct 35^{Δ} - 45^{Δ} of "A" pattern.
- F Primary and reading positions are functionally most important.

IV. Horizontal Muscle Shifts

- A. Horizontal muscles are moved in the direction that you wish to weaken them.
- B. Offsets $1/2$, $3/4$ or maximum 1 tendon with

V. Treatment of "V" Pattern Esotropia

<u>A. Collapse "V" Pattern</u>	<u>Δ of "V" Corrected</u>
IO weakening	15-25
Tuck SO OU	15-25
Vertical shift of horizontal recti	20-25
Weaken IO OU and vertical shift	25-30
SO tuck and IO weakening	40-50

- B. Horizontal surgery for deviation in primary
- C. Best procedure for "V" pattern ET
 1. If there is significant IO overaction, weaken IO's with bimedial recession or R & R.
 2. With no significant IO overaction, do bimedial recession with infraplacement.

VI. Treatment of "V" Pattern Exotropia

A. Collapse "V" Pattern Δ of "V" Corrected

IO weakening	15-25
Vertical shift of horizontal recti	15-20
IO disinsertions and vertical shift of horizontals	30

B. Horizontal surgery for deviation in primary

C. Best procedures - "V" pattern XT

1. With IO overaction, weaken IO's.
2. No IO overaction → weaken LR, with upward displacement.

VII. Treatment of "A" Pattern Esotropia

A. Collapse "A" Pattern Δ of "A" Corrected

Intrasheath SO tenotomies	30-40
Vertical shift of horizontal recti	20-25

B. Horizontal surgery for deviation in primary

C. Best procedure for "A" pattern ET

1. "A" pattern greater than 40^Δ , do SO tenotomies; horizontal surgery for residual.
2. "A" pattern less than 40^Δ , do bilateral medial recession with upward displacement; bilateral resection with downward displacement; R & R with appropriate offsets.

VIII. Treatment of "A" Pattern Exotropia

A. Collapse "A" Pattern Δ of "A" Corrected

Intrasheath SO tenotomies	30-40
Vertical shift of horizontal recti	5-20

B. Horizontal surgery for deviation in primary

C. Best procedure for "A" pattern XT

1. "A" pattern greater than 40^Δ , do SO tenotomies; horizontal surgery for residual.

2. "A" pattern less than 40^Δ , do bilateral recession with downward displacement; R & R with offsets.

D. Surgical alternatives

1. If less than 20^Δ of "A" pattern, do nothing.
2. If between 20^Δ and 40^Δ of "A" pattern or deviation in down-gaze, do bilaterals and infraplace mm or do R & R with infraplacement of LR and supraplacement of MR.
3. If 40^Δ or greater in down-gaze, do bilateral superior oblique tenotomies.

E. Indications for bilateral SO tenotomies

1. Overaction of SO OU
2. "A" pattern of at least 40^Δ in down-gaze
3. No overaction of IO's

F. Complications of SO tenotomy - reported by others

1. Urist (10 cases)
 - a. Secondary "V" pattern (4)
 - b. Unequal effects of SO (7)
 - c. Development or worsening of hyperdeviation (10)
 - d. Ptosis (3)
 - e. Change from bilateral to unilateral depression on adduction combined with unilateral elevation in adduction in the opposite eye (7)
2. Helveston (8 cases)
3. Bedrossin (10 cases)
4. Harley & Manley (20 cases)
 - a. "V" pattern (3)
5. Berke
 - a. Ptosis (1)

IX. Expected Correction from Oblique Muscle Weakening

A. SO tenotomy

- up-gaze - 0
- Primary - 10^Δ - 12^Δ

- Down-gaze - 40^Δ - 45^Δ

Amount of Eso Shift in Down-gaze with Bilateral
SO Weakening Procedure

	<u>Average</u>	<u>Maximum</u>	<u>Minimum</u>
Bedrossian	20 ^Δ	35 ^Δ	5 ^Δ
Jampolsky	45 ^Δ	65 ^Δ	35 ^Δ
Helveston	33 ^Δ	75 ^Δ	5 ^Δ
Harley & Manley	41 ^Δ	50 ^Δ	24 ^Δ
Scott, WE	43 ^Δ	55 ^Δ	30 ^Δ

B. IO weakening

- Up-gaze - 15^Δ - 25^Δ
- Primary - 0
- Down-gaze - 0

C. Summary of results of bilateral SO tenotomies done alone (10 cases)

1. Change in IO action

- a. 8 of the 20 IO (40%) changed from a pre-op underaction to a post-op overaction with an average of a +2 overaction (range +1 - +3).
- b. Two V-patterns developed; one of 8^Δ, the other 15^Δ.
- c. Complications
 - 1) 3 Brown's syndromes - one needed re-op.

* Scott WE, Jampolsky AJ, Redmond MR. Superior oblique tenotomy: Indications and complications. In Ellis FD, Helveston E (eds.): International Ophthalmology Clinics: Strabismus Surgery. Vol. 3 Boston; Little Brown Co.:1976, pp. 151-159.

X. Vertical Transposition of the Horizontal Rectus Muscles - the preferred operation in cases of A and V pattern strabismus in which oblique muscle dysfunction is inadequate to merit oblique surgery.

A. Fifty-nine patients undergoing standard horizontal surgery with half-tendon width offsets and eight patients undergoing two-thirds to full tendon width offsets were retrospectively studied.

B. Postoperative data were analyzed on a short-term (less than 6 weeks) and long-term (greater than 12 months) basis.

C. Standard horizontal surgery combined with half-tendon width vertical transposition is shown to be an effective operation for collapsing all subgroups of A and V pattern strabismus when indications are appropriate.

D. The initial correction to within $\pm 10^\Delta$ of pattern was 96% over all, with 78% remaining collapsed to within $\pm 10^\Delta$, over an average 36-month follow-up.

* Scott WE, Drummond GT, Keech RV. Vertical offsets of horizontal recti muscles in the management of A and V pattern strabismus. Aust NZ J Ophthalmol 1989;17(3):281-288.

Vertical Deviations

I. Differential Diagnosis

- A. Superior Oblique Palsy - most common cause of vertical deviation. Assume a superior oblique palsy until proven otherwise
- B. Monocular Elevation Deficiency
- C. Inferior Oblique Palsy
- D. Brown's Tendon Sheath syndrome
- E. Orbital floor fractures with inferior rectus entrapment
- F. Grave's Ophthalmopathy
- G. Myasthenia Gravis
- H. Progressive External Ophthalmoplegia

II. Superior Oblique Palsy

A. Etiology

- 1. Congenital
- 2. Acquired - usually following closed head trauma

B. Clinical features

- 1. Hypertropia or hypotropia - determined by fixing eye.
- 2. Abnormal head positions.
 - a. Head tilt to opposite side most common; i.e., left tilt for right superior oblique palsy.
 - b. Patients occasionally adopt a face turn to the same side as the head tilt.
- 3. Amblyopia- uncommon
- 4. May be unilateral or bilateral

Diagnostic Criteria

Unilateral

- Hypertropia in primary
- Unilateral oblique dysfunction
- Positive 3-step test
- Hyper worse on head tilt to same side as palsy
- Excyclotorsion $<12^\circ$

Bilateral

- May or may not have hyper in primary
- Bilateral oblique dysfunction
- Reversing hypers on side gazes and head tilts
- Excyclotorsion $>12^\circ$
- V pattern

5. Excyclotorsion (see photos: [link](#))

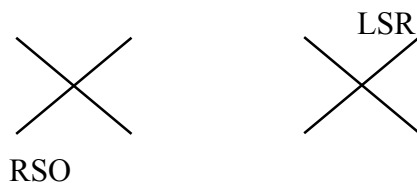
- a. Measured subjectively with the Double Maddox Rod (in primary and reading positions).
- b. Measured objectively with indirect ophthalmoscopy, fovea should be adjacent to the lower third of the disc.

C. Parks' three step test

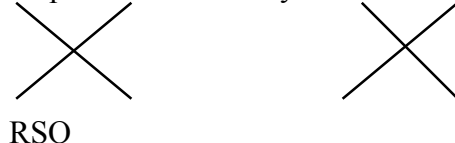
1. Step 1 - hypertropia in primary position; i.e.: RHT - secondary to weak depressors OD or weak elevators OS.



2. Step 2 - hyper increases in one lateral gaze. RHT increase in left gaze - indicates weak RSO or LSR.



3. Step 3 - Bielschowsky Head Tilt Test; RHT increases on right tilt - RSO is paretic.



If the right superior oblique is paretic the right superior rectus is acting against decreased opposition to elevation thus RHT increases on right tilt.

*Parks MM. Isolated cyclovertical muscle palsy. Arch Ophthalmol 1958;60:1027-1035.

D. The diagnosis of a superior oblique palsy is made by the 3-step test and not on the actions of the oblique muscles seen on versions.

E. Surgical treatment of superior oblique palsy - operations of choice

1. When antagonist inferior oblique is overacting and deviation in primary is not greater than 13^{Δ} , recess the inferior oblique.
 - a. The amount of deviation corrected in primary is dependent on the amount of overaction of the inferior oblique
 - b. Doesn't correct any vertical deviation outside it's field of action
 - i. Any deviation outside the field of action, should be corrected with an additional muscle.
2. If deviation is greater than 13^{Δ} with an overacting antagonist inferior oblique, recess inferior oblique and contralateral inferior rectus
3. If antagonist inferior oblique is not overacting, recess contralateral inferior rectus.
4. Rule of Thumb: 1mm recession = 3^{Δ} of correction.
5. If forced ductions show a tight superior rectus on the side of the deviation, recess it instead of the contralateral inferior rectus.
6. If deviation is greater than 35^{Δ} think about doing 3 muscles.
7. Superior oblique tucks
 - a. Best in bilateral cases.
 - b. Help torsion.
 - c. Correct a small amount of vertical in primary.
 - d. May cause a limitation in elevation in adduction (iatrogenic Brown's).
 - e. Correct ET in down-gaze.
8. Harada-Ito procedure (anterior and temporal displacement of anterior fibers of the superior oblique) is an alternative to the superior oblique tuck.
 - a. Corrects torsion.
 - b. May be combined with other rectus muscle surgery.
 - c. To correction torsion move tendon more laterally
 - d. To correct more esotropia in downgaze, move the tendon posteriorly up to 8 mm along the lateral rectus. This can correct up to 15 PD in downgaze.

F. Superior oblique palsies - surgical cases

1. 118 cases at University of Iowa Hospitals, 1972-1983
 - a. 84 (71.1%) unilateral, subdivided into 8 patterns
 - b. 26 (22.1%) bilateral, subdivided into 4 patterns
 - c. 8 (6.8%) "Masked" bilateral

G. A simplified unilateral superior oblique palsy approach

1. Steps in the management of unilateral superior oblique palsy
 - a. Measure deviation and grade versions - should coincide.
 - b. One muscle surgery - $<13^\Delta$ in primary.
 - c. If oblique done
 - 1) No deviation out of field of action.
 - 2) Amount of deviation is proportional to amount of overacting.
 - 3) Do IO if overacting.
 - 4) If IO normal, and SO is UA tuck SO. Deviation $>$ in field of action of SO.
 - 5) If there is a spread of comitance, do vertical rectus.
 - 6) If "fallen eye", relieve restriction - recess IR.
 - d. Two muscle surgery
 - 1) Deviation between 13^Δ - 30^Δ .
 - 2) If significant IOOA - weaken.
 - 3) If normal inferior oblique and SO underaction - SO tuck.
 - e. Other procedures
 - 1) Deviation greater in upgaze - ipsilateral SR.
 - 2) Deviation in downgaze - yoke IR.
 - 3) Spread of comitance - vertical recess/resect.
 - f. Deviation greater than 30^Δ - refer to someone you don't like

- * Scott WE, Kraft SP. Classification and surgical treatment of superior oblique palsies: I. Unilateral superior oblique palsies. In: Pediatric Ophthalmology and Strabismus: Transactions of the New Orleans Academy of Ophthalmology. New York;Raven Press:1986, pp. 15-38.
- * Scott WE, Kraft SP. Classification and treatment of superior oblique palsies: II. Bilateral superior oblique palsies. In: Pediatric Ophthalmology and Strabismus: Transactions of the New Orleans Academy of Ophthalmology. New York;Raven Press:1986, pp. 265-291.
- * Knapp P. Classification and treatment of superior oblique palsy. Amer Orthop J 1974;24:18-22.

III. Monocular Elevation Deficiency

A. Three types of monocular elevation deficiency

1. Inferior rectus restriction.
 - a. Positive forced ductions.
 - b. Normal forced generations.
 - c. Normal saccades of superior rectus.
2. Elevation weakness

- a. Etiology - palsy of the superior rectus and inferior oblique secondary to a supranuclear disturbance.
 - b. Free forced ductions.
 - c. Reduced forced generations.
 - d. Reduced saccadic velocities.
- 3. Combination
 - a. Positive forced ductions.
 - b. Reduced forced generations.
 - c. Reduced saccadic velocities.

B. Clinical features

- 1. Hypotropia
- 2. Amblyopia
- 3. Ptosis - true and pseudo
- 4. Chin up head position - if abnormal head position is not present amblyopia is likely.
- 5. May have restriction of inferior rectus.
 - a. Look for lower lid crease - the crease becomes more pronounced on attempted upgaze if IR restriction is present.
 - b. Bell's phenomenon should be normal unless the IR is restricted.
 - c. Duction = version, if IR restriction is present.
 - d. Forced ductions are positive to elevation.

C. Indications for treatment

- 1. Large vertical deviation with ptosis
- 2. Abnormal head position

D. Surgical treatment

- 1. Relieve restriction if present
- 2. Transposition procedure
 - a. If there is no restriction of IR.
 - b. If there is a residual hypotropia after inferior rectus recession.
- 3. Type I - recess inferior rectus
- 4. Type II - transpose medial rectus and lateral rectus superiorly (Knapp procedure)
- 5. Type III - recess inferior rectus and transpose medial and lateral rectus

6. A Knapp procedure will correct approximately 35^Δ of deviation in patients without inferior rectus restriction.

* Scott WE, Jackson OB. Double elevator palsy: the significance of inferior rectus restriction. Amer Orthop J 1977;27:5-10.

IV. Inferior Oblique Palsy

A. Clinical features

1. Hypertropia or hypotropia - determined by fixing eye
2. Positive 3-Step test
3. Overaction of ipsilateral superior oblique
4. Duction better than version
5. Free forced ductions
6. Abnormal head position
7. Pupils are normal
8. Usually due to direct orbital trauma or viral illness

B. Indications for treatment

1. Large vertical deviation
2. Abnormal head position
3. Diplopia

C. Treatment

1. Superior oblique intrasheath tenotomy
2. Normalizes the action of the superior oblique
3. Superior oblique z tenotomy and tendon spacers have not given enough effect.

* Scott WE, Nankin SJ. Isolated inferior oblique paresis. Arch Ophthalmol 1977;95:1586-1593.

V. Brown's Syndrome

A. Initially described by Dr. Harold Whaley Brown in 1944; for many years known as the "superior oblique tendon sheath syndrome", a misnomer

B. Clinical features

1. Must have
 - a. Deficient elevation in adduction.
 - b. Less elevation deficiency in midline.
 - c. Minimal or no elevation deficit in abduction.
 - d. Minimal or no superior oblique overaction.
 - e. Divergence in upgaze producing a "V" pattern.
 - f. Positive forced duction testing (accentuated by retroplacement of the globe).
2. May also have
 - a. Downshoot in adduction.
 - b. Widened palpebral fissure on adduction.
 - c. Anomalous head posture (chin up or face turn away from the affected eye).
 - d. Primary position hypotropia.
 - e. Tenderness to palpation over the trochlea.
 - f. "Click" or snapping sensation in attempted elevation in adduction.
 - g. Palpable nodule.

C. Classification

1. Brown initially divided cases into "true" and "simulated" - not useful, since the etiology upon which the subdivisions were based is incorrect.
2. Essentially there are two major forms:
 - a. Congenital.
 - b. Acquired.
 - c. Either form can be intermittent.

D. Heredity

1. Autosomal dominant with incomplete penetrance and variable expressivity
2. Most cases are sporadic

E. Etiology

1. Anatomic considerations
 - a. The tendon and Tenon's capsule, through which it passes, create a sleeve arrangement, permitting easy movement of the tendon.

- b. The intermuscular septum envelops the superior oblique tendon and maintains connections with the superior rectus.
 - c. Each fiber of the superior oblique tendon acts independently as a cord from the muscle fibers to the insertion thus producing a telescoping or slide-by fashion of movement.
 - d. A bursa-like structure is found between the tendon and the trochlear saddle.
 - e. The intratrochlear superior oblique tendon has a highly vascular sheath.
- 2. Probable final common pathway: defect in the trochlea/tendon complex
 - a. Any interference with the normal telescoping movement of the tendon
 - b. Excess fluid accumulation or concretions in the bursa
 - c. Distension of the vascular sheath
 - d. Stenosing tenosynovitis
 - 1) Movement of the tendon in the trochlea creates a metabolic requirement for repairing "wear and tear", which is normally performed by the vascular intratrochlear sheath.
 - 2) If the wear and tear is excessive, thickening and stenosis of the sheath occurs with secondary enlargement of the tendon at the point of restricted gliding.
 - 3) This may be very similar to trigger finger (digital tenosynovitis stenosis) and congenital trigger thumb.
 - e. Inflammation around the trochlea
 - 1) Associated with juvenile and adult rheumatoid arthritis.
 - 2) Digital pressure over the trochlea can produce pain, but helps to relieve the restriction.
 - 3) Sudden release (spontaneous resolution) or attempt to move the superior oblique tendon is often accompanied by discomfort localized to the trochlear area.
 - 4) Can occasionally see thickening and edema by CT.
 - 5) Improvement with injection of steroids (even in non-rheumatoid cases).
 - f. Oblique muscle palsy, Brown's syndrome, or both
 - g. Postoperatively: after superior oblique, orbital, retinal, or sinus surgery.
 - h. Conclusions
 - 1) Idiopathic cases of Brown's syndrome may be the result of inflammation caused by wear and tear, in the setting of a congenital anomaly or predisposition.
 - 2) Inflammation may subside, leaving persistent adhesions; therefore, absence of trochlear tenderness or swelling does not exclude active stenosing tenosynovitis.
 - 3) Intermittent "click" variety may represent an intermediate stage in the resolution of the constant variety.
 - 4) Congenital and acquired Brown's syndrome may be a continuum.

F. Treatment

- 1. Non-surgical treatment
 - a. Observation: spontaneous regression is common in acquired and intermittent cases, but less common in congenital and constant cases.
 - b. Elevation in adduction exercises.
 - c. Steroid injections into the superonasal quadrant (paratrochlear) - 40mg Depo-Medrol, may need to be repeated at monthly intervals.

d. Treat the underlying condition, e.g. JRA.

2. Surgical treatment

a. Indications:

- 1) Primary position hypotropia and/or abnormal head position.
- 2) Occasionally, cosmetically unacceptable downshoot of the eye in adduction.

b. Tenotomy is the current procedure of choice

- 1) Does not typically lead to torsional difficulties or complete superior oblique paralysis.
- 2) The intermuscular septum serves as an insertion for the proximal end of the cut tendon, transmitting the superior oblique force to the distal severed tendon.
- 3) Some advocate inferior oblique recession to decrease the incidence of post-op superior oblique palsy - this is still controversial.

c. Surgery at the trochlea (the supposed site of abnormality) is fraught with complications.

d. Silicone tendon expanders.

* Sprunger DT, von Noorden GK, Helveston EM. Surgical results in Brown's syndrome. J Ped Ophthalmol Strab 1991;28:164-167.

* Wilson ME, Eustis HS, Parks MM. Brown's syndrome. Surv Ophthalmol 1989;34:153-172.

* Scott WE, Arthur BW. Current approaches to superior oblique muscle surgery. Focal Points 1988: Clinical Modules for Ophthalmologists Vol. 4, Module 3, 1988.

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

VI. Blow-out Fractures

A. Types

1. Isolated floor fractures
2. Tripod fracture (dislocation fracture of the zygoma)
3. Complex facial fracture with orbital floor fracture
4. Infra-orbital rim fracture with orbital floor fracture
5. Medial wall involvement with orbital floor fracture

B. Etiology

1. Auto accidents

2. Fist injury
3. Sports
4. Blunt hurled objects
5. Falls

C. Signs and symptoms

1. Ecchymosis
2. Diplopia
3. Other ocular damage
4. Paresthesia of infra-orbital area
5. Enophthalmos
6. Abnormal head position

D. Diagnosis

1. Motility exam - measurements in all positions
2. Forced ductions
3. Measure intraocular pressure in upgaze and primary
4. Saccadic velocities
5. Radiographs
 - a. Regular films.
 - b. Tomographs.
6. Complete examination
7. ENT consultation

E. Motility problems to consider

1. Entrapment of inferior rectus or inferior oblique
 - a. Hypotropia in primary.
 - b. Hypotropia increases in upgaze.
 - c. Positive forced ductions.
 - d. Once edema has subsided, operate to free entrapped muscle.

2. Paresis of inferior rectus
 - a. Etiology - trauma to nerve to IR either at the time of injury or at the time of repair of floor.
 - b. If paresis exists without entrapment, hyper in primary.
 - c. If paresis exists with entrapment, ortho to slightly hypotropic or hypertropic in primary.
 - d. May recover with time.
 - e. Important to recognize.

3. Entrapment of medial rectus

4. Paresis of medial rectus

F. Other ocular damage

1. Corneal problems
2. Hyphema
3. IR palsy
4. Pupil injury
5. Cataracts
6. Vitreous hemorrhage
7. Choroidal rupture
8. Optic nerve damage
9. Retinal problems - hemorrhage, edema

* Leibsohn J, Burton TC, Scott WE. Orbital floor fractures: a retrospective study. Ann Ophthalmol 1976;8:1057-1062.

VII. III Nerve Palsy

A. Congenital

1. Etiology
 - a. Perinatal trauma causing injury to the peripheral nerve.
 - b. Familial (rare; one case of a daughter and father with unilateral double elevator palsy, hypotropia, ptosis).

- c. Developmental defect of nucleus (may have other neurologic abnormalities) or motor fiber portion of third nerve complex.
 - d. Rare causes.
 - 1) Ophthalmoplegic migraine
 - Usually in children
 - Positive family history of migraine
 - Headache, nausea, vomiting resolved with onset of paresis
 - Paresis usually improves within one month; occasionally permanent
 - 2) Cyclic (see XII).
2. Differential diagnosis
- a. Congenital Horner's syndrome (because of aberrant reinnervation with pupillary miosis; ptosis).
3. Characteristics
- a. Exotropia, hypotropia (if fixing with nonparetic eye), hypertropia (if fixing with paretic eye).
 - b. Intact pupillary light and accommodation responses; may have aberrant regeneration with pupillary constriction on adduction.
 - c. Ptosis.
 - d. Limitation of elevation, depression, adduction.
 - e. May have MR function.
4. Management (See C.)

B. Acquired

1. Etiology
- a. Brainstem lesion (extremely rare, often produce bilateral defects. e.g. encephalitis, metastases, ischemia).
 - b. Inflammation (encephalitis, meningitis secondary to tuberculosis, varicella, herpes zoster, other infectious causes, idiopathic intracranial hypertension, giant cell arteritis, toxins causing polyneuritis).
 - c. Vascular lesion (aneurysm, ischemia associated with diabetes, hypertension, atherosclerosis, migraine).
 - d. Neoplasm (metastases, lymphoma, leukemia, meningiomas, pituitary tumors, craniopharyngiomas, nasopharyngeal tumors).
 - e. Demyelinating disease.
 - f. Trauma (causing contusion, stretching).
 - g. Miscellaneous (leukemia, polyarteritis nodosa, sarcoid, myasthenia gravis, etc.).
 - h. Most common: neoplasm, aneurysm, ischemia, and trauma.
2. Differential diagnosis
- a. Myasthenia gravis (can mimic pupil-sparing IIIrd nerve palsy).
3. Characteristics
- a. Partial or complete.

- b. Pupil-sparing suggests ischemia (most recover by 3-6 months); pupil involvement suggests compressive lesion .
- c. Exotropia, hypotropia.
- d. Ptosis.
- e. Limitation of elevation, depression, adduction.
- f. May develop aberrant regeneration.
 - 1) Lid-gaze dyskinesia (retraction on attempted down-gaze or adduction, narrowing of fissure on abduction).
 - 2) Pupil gaze dyskinesia (more pupil constriction on convergence; constriction on attempted downgaze).
 - 3) Globe retraction on attempted vertical gaze.
 - 4) Adduction on attempted vertical gaze.

C. Treatment

- 1. Superior oblique tenotomy; maximal LR recession & MR resection
- 2. Combined recess/resect procedure with transposition to improve the weakest ocular rotation
- 3. Transposition of insertion of SO tendon to 2-3mm anterior to medial side of SR insertion; large LR recession \pm MR resection
- 4. For ptosis: - frontalis suspension
 - if aberrant regeneration with lid retraction on adduction, try horizontal R & R on unaffected eye

* Buckley EG, Townshend LM. A simple transposition for complicated strabismus. Amer J Ophthalmol 1991;111:302-206.

* Gottlob I, Catalano RA, Reinecke RD. Surgical management of oculomotor nerve palsy. Amer J Ophthalmol 1991;111:71-76.

* O'Donnell FE, Del Monte M, Guyton DL. Simultaneous correction of blepharoptosis and exotropia in aberrant regeneration of the oculomotor nerve by strabismus surgery. Ophthalmic Surg 1980;11(10):695-697.

VIII. Congenital Fibrosis Syndrome (Crawford, JL; Apt L, Axelrod RN)

Synonyms: General Fibrosis syndrome, Congenital Ophthalmoplegia, Strabismus fixus.

A. Autosomal dominant or sporadic - 60% have positive family history

B. Characteristics

1. Head tilt - chin up 16/16.
2. Inability to elevate or depress eye 16/16.
3. Severe blepharoptosis 16/16.
4. Convergent jerky movements on attempted elevation.
5. Horizontal movements severely restricted 16/16, exotropic 8/16, esotropic 1/16.
6. Hyperopic astigmatism 11/14.
7. Bilateral decreased visual acuity 16/16.
8. Present at birth.
9. Reported associations: Coloboma, Retinitis Pigmentosa.

C. Pathology

1. Posterior membranous insertion of recti and anterior and medial insertion of obliques.
2. Fibrosis of muscle and Tenon's.
3. Adhesions between muscles, Tenon's & globe.
4. Inelastic & fragile conjunctiva.

D. Require multiple procedures for horizontal, vertical strabismus and later ptosis repair

IX. Graves' Ophthalmopathy

A. Incidence of Graves' at the University of Iowa - approximately 175 patients seen in an 8 year period.

B. Eye changes in Graves' disease

1. No signs or symptoms
2. Only signs, no symptoms - upper lid retraction, stare, with or without lid lag and proptosis
3. Soft tissue involvement
4. Proptosis
5. Optic nerve involvement

C. Patient characteristics of 25 patients studied with Graves'

1. Proptosis 20/25 (80%)

2. Lid involvement 18/25 (72%)
3. Corneal involvement 14/25 (56%)
4. Soft tissue involvement 9/25 (36%)
5. Optic nerve involvement 7/25 (28%)

D. Thyroid Function - 25 patients

1. Laboratory diagnosis of hyperthyroidism 23/25
2. Demonstrated symptoms consistent with thyroid dysfunction 2/25
3. Received some form of medical treatment for their thyroid condition, i.e., surgery, I131, medication 21/25

E. Thyroid state at time of eye muscle surgery - 25 patients

1. Hypothyroid on supplemental treatment 50%
2. Euthyroid 50%
3. Hyperthyroid 0%

F. Pre-op evaluation

1. Document other ocular involvement
 - a. Usual eye exam.
 - b. Visual fields.
 - c. Iris angiogram.
 - d. Diplopia fields.
 - e. Intraocular pressures-in primary, upgaze and downgaze.
 - f. Echography.
 - g. Optic nerve evaluation.
 - 1) Baseline VER.
 - 2) Farnsworth-Munsell 100 hue.
 - 3) Flicker Fusion Frequency.
 - h. If esotropia improves in downgaze, the esotropia in primary position is likely the result of tight inferior rectus muscles

G. Muscle involvement - 25 patients

1. Inferior rectus 20/25 (80%)
2. Medial rectus 11/25 (44%)

3. Combined inferior rectus and medial rectus 9/25 (36%)
4. Superior rectus 6/25 (24%)
5. Lateral rectus 0/25

H. Pre-op treatment - 25 patients

1. Fresnel prisms 12/25 (48%)
2. Patched one eye 6/25 (24%)
3. No treatment - able to ignore diplopia 7/25 (28%)

I. Treatment - 25 patients

1. Elected to have surgery 22/25
2. Prior orbital decompressions 5/22

J. Surgical technique

1. Tight muscles, therefore large amounts of surgery are necessary
2. Adjustable sutures
3. Lower lid lag prevented by cleaning IR 14-16 mm back

K. Surgical procedures - 22 patients

1. 1 muscle - IR recession 10/22
SR recession 2/22
2. 2 muscle - Bilateral IR recessions 1/22
SR recession, MR recession 1/22
IR recession, LR resection 1/22
Bimedial recessions 1/22
3. 3 muscle - IR recession, bimedial recessions 2/22
LR resection, bimedial recessions 1/22
4. 4 muscle - Bilateral IR recessions, bimedial recessions 2/2
Bimedial recessions, IR recession, SR recession 1/22

L. Surgical amounts

1. IR recession (3mm - 7 mm)

2. MR recession (3.5mm - 8.5mm)

3. SR recession (4 mm - 5mm)

M. Post-op results - 22 patients

1. Able to fuse in primary position without prisms or abnormal head position 18/22

2. Fused in primary position with the aid of 8^Δ vertical prism 1/22

3. Unable to fuse but ignored the second image 3/22

N. Post-op motility - 22 patients

1. Limitation of upgaze 10/22 (45%)

2. Limitation of abduction 10/22 (45%)

3. Limitation of downgaze 7/22 (32%)

4. Limitation of adduction 2/22 (9%)

5. Normal versions 4/22 (18%)

O. Post-op complications

1. Down-gaze problems

2. "A" pattern- if weaken both inferior rectus muscles and both medial rectus muscles, the superior oblique muscles may overact

3. Anterior segment ischemia

4. Instability

P. Secondary procedures

1. Recess antagonists on adjustable

2. Advance previously recessed muscle

3. Further recession IR

4. Recess contralateral IR

* Scott WE, Thalacker JA. Diagnosis and treatment of thyroid myopathy. Ophthalmol 1981;88:493-498.

* Lueder GT, Scott WE, Kutschke PJ, Keech RV. Long-term results of adjustable suture surgery for strabismus secondary to thyroid ophthalmopathy. Ophthalmol 1992;99(6):993-997.

Q. 55 patients

47 had adjustable suture strabismus surgery.

8 managed with Fresnel prisms alone.

R. Average follow-up = 41 months (range, 6 to 168 months)

S. 47 surgical patients

40 (85%) - 1 procedure

6 - 2 procedures

1 - 4 procedures

T. Surgery

1. 56 total procedures

22 one-muscle

13 two-muscle

14 three-muscle

7 four-muscle

2. 117 total muscles operated

53 inferior rectus

50 medial rectus

12 superior rectus

2 lateral rectus

3. 37 (66%) patients underwent postoperative adjustment

4. Complications

1 case of anterior segment ischemia

13 cases of lower eyelid retraction

U. Results after initial surgery

1. Excellent 22 (47%)

2. Good 12 (26%)

2 small abnormal head position

1 rare diplopia in downgaze

9 required prisms in glasses

3. Fair or Poor 18 (38%)

6 progressive restriction of other extraocular muscles

5 unrecognized superior rectus involvement

- 4 A-pattern exotropia
 - 1 intermittent exotropia in reading position
 - 1 overcorrection
 - 1 small intermittent esotropia
 - 7 had additional surgery with 5 obtaining good or excellent results

- V. 24 patients initially given prisms
 - 16 required surgery
 - 8 managed exclusively with prisms

X. Chronic Progressive External Ophthalmoplegia

A. Characteristics

- 1. 50% autosomal dominant
- 2. Average age of onset - 23 years
- 3. Usually commences with ptosis
- 4. Slowly progressive palsy of all extraocular muscles
- 5. Gaze function primarily involved rather than individual extraocular muscle
- 6. Upward gaze and convergence involved first, then lateral gaze
- 7. Diplopia unusual
- 8. Systemic muscular involvement, primarily head, neck and upper limbs

B. Etiology

- 1. Prior to 1951 - nuclear atrophy
- 2. 1951 (Kiloh and Nevin) - myopathy
- 3. 1969 (Daroff) - myopathy vs. CNS brain stem disease

XI. Myasthenia Gravis

A. Signs and symptoms

- 1. Ptosis and diplopia are the most common presenting symptoms
 - Diplopia may be present without ptosis
 - a. Characteristically signs and symptoms fluctuate, often ptosis is unilateral or bilateral and asymmetric.

- b. One lid may retract on attempted elevation of other lid.
- c. Cogan's lid twitch (temporary lid elevation in redirection of gaze from downward to primary).
- d. Ptosis may worsen with prolonged upgaze or sidegaze and improves after rest.
- e. Extraocular muscle involvement: the medial rectus and elevators are commonly involved but involvement may mimic any isolated nerve or muscle palsy, internuclear ophthalmoplegia, gaze palsy, or double elevator palsy.

B. Diagnostic tests

- 1. Tensilon: must have a good endpoint.
- 2. Prostigmine: especially useful for
 - a. Children who won't cooperate for IV placement.
 - b. Adults in whom one wishes to measure prism diopters of deviation before and after drug given.
 - c. Adults with equivocal Tensilon tests.
- 3. Harvey-Maslan Repetitive Supramaximal motor stimulation.
- 4. Acetylcholine receptor antibody testing

C. Prognosis and treatment of diplopia

- 1. 50%-80% of ocular myasthenia will go on to develop generalized myasthenia, usually within two years
- 2. Diplopia often responds poorly to anticholinesterase therapy and may do better with Prednisone
- 3. Diplopia fluctuates, so prisms or surgery are limited to use after the disease has stabilized in the patients with residual deviation

XII. Cyclic Third Nerve Palsy

A. Characteristics - 60 cases reported in world literature

- 1. Paralysis of the third nerve with alternating paretic and spastic phases
- 2. Usually congenital or onset in first few months of life
- 3. Usually no history of trauma
- 4. Negative family history
- 5. Remains throughout life

B. Clinical Findings

1. Paralysis is usually complete
2. Degree of involvement
 - a. Pupillary musculature uniformly involved.
 - b. Ciliary body most likely involved in all.
 - c. Levator and medial rectus are the most common extra-ocular muscles showing cycling.
3. Pupils
 - a. During the paretic phase, no direct or consensual light reaction.
 - b. Atropine dilates the pupil and interrupts cycling phenomenon.
 - c. Pilocarpine produces miosis and abolishes cycling.
 - d. Cocaine dilates the pupil but does not interrupt the cycling.

C. Cycling phenomenon - constant time intervals for a given patient.

1. Paretic phase
 - a. Complete ptosis.
 - b. Pupil dilated and fixed.
 - c. Eye exotropic and hypotropic.
 - d. Accommodation relaxed.
2. Spastic phase
 - a. Lid elevates.
 - b. Pupil constricts.
 - c. Eye returns to midline.

3. Each phase lasts 30-60 seconds

D. Convergence or adduction of the involved eye tends to prolong the spastic phase while abduction tends to shorten it.

E. Cycles usually continue during sleep, but intervals are lengthened, with shortening of the spastic phase

F. Cycles are abolished under anesthesia

G. Etiology - all speculative, no autopsy material

1. Fuchs and Bielschowsky - rhythmic variations in blood supply to the third nerve nucleus
2. Axenfeld and Schurenberg - lesion peripheral in third nerve; cycling due to intermittent pressure on the nerve

3. Behr and Bielschowsky - partial degeneration of third nerve nucleus; remaining ganglion cells of the nucleus respond to rhythmic impulses, which reach the partially destroyed nucleus from higher, presumably vegetative, centers
 4. Stevens - reported a 25 year old female
 - developed a cyclic third nerve palsy which later became complete.
 - 14 months later papilledema was noted.
 - Dx - brain stem glioma.
 - Rx - radiation
 - Only known case of acquired cyclic palsy.
- * Clarke WN, Scott WE. Cyclic third nerve palsy. A report of two cases. J Ped Ophthalmol Strab 1975;12(2):94-99.
- * Loewenfeld IE, Thompson HS. Oculomotor paresis with cyclic spasms. A critical review of the literature and a new case. Surv Ophthalmol 1975;20(2):81-124.

Nystagmus

I. Spasmus Nutans

A. A clinical entity consisting of the triad of nystagmus associated with head nodding and abnormal head positions

B. Nystagmus and head nodding are usually present together but either may occur alone

C. Raudnitz(1897) - in a series of 47 cases found

- head nodding 87%
- nystagmus 80%
- abnormal head position 38%

D. Most often nystagmus is very fine, rapid, pendular, but may be rotary or vertical

1. Asymmetric in the two eyes
2. Often varies in different positions of gaze
3. May be monocular, spasmus nutans is the most common cause of unilateral nystagmus in infancy
4. EOG studies confirm rapid frequency (3-10 HZ), pendular waveform, marked asymmetry between eyes, highly variable frequency (changing from sec. to sec.)
subclinical nystagmus may be diagnosed by EOG

E. Head nodding

1. Intermittent and irregular; may be horizontal, vertical or both
2. Head movements are not compensatory for nystagmus
3. Disappears during sleep or when eyes are closed

F. Abnormal head position

1. Turning or tilting of the head
2. Disappears during sleep
3. Never the only sign of spasmus nutans

G. Age of onset usually 4-12 months but ranges from 6 weeks to 3 years

H. Variable duration, weeks - months. In Hermann's study of 20 cases all recovered within 12 months, but Norton and Cogan⁴⁴ report 2 cases which lasted for 8 years

I. Pathogenesis - unknown. Get CT

J. No treatment

K. Prognosis is good, seems to be a self limiting entity

L. Associated conditions

1. Strabismus - 7 of 20 cases reported by Norton and Cogan had a heterotropia
2. Refractive errors
3. No consistently associated neurological conditions

* Farmer J, Hoyt CS. Monocular nystagmus in infancy and early childhood. Amer J Ophthalmol 1984;98:504-509.

M. May mimic life threatening intracerebral tumor

1. Lavey et. al. reported on 20 cases of early acquired nystagmus, which was the first sign of intracerebral glioma
2. 16/20 onset of nystagmus prior to 1 year
3. 10/20 originally diagnosed as spasmus nutans. Average delay in correct diagnosis of 14.5 months
4. 16/20 originally unilateral nystagmus and 11/20 had head nodding and/or torticollis
5. Associated signs, which necessitate CT scan but which, did not present initially
 - a. Optic nerve atrophy developed eventually 18/20.
 - b. Diencephalic syndrome - emaciation despite food intake, hyperactivity and euphoria, skin pallor, hypotension, hypoglycemia, neuroendocrine disturbances 7/20.
 - c. Papilledema and/or increased head circumference due to obstructive hydrocephalus 9/20.
 - d. Afferent pupillary defect or loss of fixation ability.
6. Onset of nystagmus after 1 year should have CT
7. Tumor is low grade astrocytoma involving chiasm, hypothalamus and optic nerve
8. R/4500-5000 rads. Good prognosis for life but optic atrophy progresses in most.

9. There is no definite sign to differentiate the acquired nystagmus of spasmus nutans from intracerebral glioma in some reported cases. Therefore, careful consideration of enhanced CT scan in all cases of acquired nystagmus of undetermined etiology is warranted.

* Lavey MA, O'Neill JF, Chu FC. Acquired nystagmus in early childhood: A presenting sign of intracranial tumor. *Ophthalmol* 1984;91:425-435.

II. Pendular Nystagmus

- A. Oscillations that in some positions of gaze are approximately equal in rate in both directions.
- B. Almost always horizontal.
- C. Absence of central vision causes loss of fixation reflex.
- D. 2-4-6 Rule:
 - If visual loss happens before 2 years of age nystagmus results.
 - 2-6 years - irregular and unsustained movements of fixation but not a true nystagmus.
 - Older than 6 years - no abnormal eye movements.
- E. Patients may attempt to compensate with synchronous, contraversive head-eye movements.
- F. Nystagmus disappears in sleep and with barbiturates.
- G. Etiology - any process causing loss of central vision early in life.
- H. Associated findings
 1. Strabismus 6%
 2. Refractive errors - often CHA.
 3. OKN response (Cogan)
 - Type I - good OKN.
 - Type II - no OKN, poorer prognosis, repeat ocular exam.

III. Latent Nystagmus

- A. Characteristics
 1. Bilateral jerk nystagmus
 2. Fast phase towards the fixing eye

3. Occurs when light stimulus to one eye is diminished or when one eye covered
4. Usually congenital
5. May occur in conjunction with manifest nystagmus
 - i.e., nystagmus amplitude increases with occlusion of one eye

B. Etiology

Thought to be the result of the difference in the quality of retinal images.

C. Methods of visual acuity assessment

1. Monocular
 - a. High plus lens occluder
 - Blurs VA of one eye without diminishing light stimulation to decrease the intensity of the latent nystagmus.
 - Problems.
 - * Patients may peek around the lens.
 - * May have such a strong eye dominance that they continue to fixate with the occluded eye, thus the VA of the non-dominant eye cannot be assessed.
 - b. Vectograph
 - Patient wears polarized lenses (not as dissociative as an occluder or patch).
 - Slides.
 - 2 polarized to test OD
 - 2 polarized to test OS
 - Polarized to test binocularly
 - Problems.
 - * May suppress one eye and therefore cannot see letters when attempting to assess VA of the suppressing eye.
 - c. Duochrome slide with green filter occluder
 - Duochrome superimposed over Snellen letters or numbers (less dissociative than occluder or patch).
 - Patient instructed to read letters/numbers on the red background with the unoccluded eye.
 - Problems.
 - * Wavelength of the green filter must be the same as the Duochrome slide to render letters on the red background invisible
 - * May suppress one eye, making VA assessment of that eye impossible
 - d. Vertical prism
 - Significantly separates distance between chart viewed with each eye while at the same time blurring the image of the eye under the prism
 - Problems
 - * May prefer to fix with eye occluded with vertical prism

2. Binocular
 - a. Always assess for best VA.

D. Other Helpful Hints

1. Abnormal head positions (AHP)
 - a. Always allow the use of an AHP.
 - b. May adopt an AHP for null point.
 - c. May adopt a face turn to place the fixing eye in adduction to dampen nystagmus.
2. Near vs. distance visual acuity
 - a. Near VA is often better than distance VA as nystagmus is dampened with accommodative convergence

- E. Amblyopia should be treated in the traditional fashion.
Patients will often adopt a face turn to fix in adduction and dampen nystagmus.

IV. Congenital Nystagmus

- A. Pendular nystagmus which converts to a jerk type on gaze to either side
- B. Usually horizontal
- C. Always bilateral
- D. Head movements opposite to direction of nystagmus
- E. Etiology - unknown
- F. Visual acuity
 1. Usually in 20/40 to 20/80 range
 2. Reported as good as 20/30
 3. Better when measured binocularly due to a latent nystagmus which is often superimposed
 4. Convergence has a dampening effect on the nystagmus
 5. Near vision often much better
 6. 80% can read normal print books and attend normal schools
- G. "Quiet Zone"

1. Position of eyes where amplitude of oscillation is decreased
2. Often at extremes of lateral gaze
3. Child uses head turn to facilitate gaze straight ahead while maintaining eyes in quiet zone
4. May show accompanying chin elevation or depression

H. Cogan's sign- vertical optokinetic nystagmus responses suggest better vision and not a sensory cause

I. Medical treatment

1. Quiet the nystagmus
 - a. Minus lenses.
 - b. Cycloplegic drugs.
 - c. Base out prism.
2. Prism realignment
 - a. Stimulates a vergence movement.
 - b. Cosmetically poor.
 - c. Prism distortion.

J. Surgical treatment

1. Objective - to mechanically move the quiet zone to a cosmetically more acceptable position and relieve head turn

K. Surgical technique

1. Parks' "Straight Flush" technique -
 - a. R & R 5, 6, 7, 8 mm.
 - b. ET eye 5 & 8 mm (total 13).
 - c. XT eye 6 & 7 mm (total 13).
2. Works well for head positions of 30° or less.

V. Retrospective review - 1974-1982

A. 32 patients with congenital nystagmus

1. Age range 4-20 years
2. 22 males, 10 females

3. Follow-up 6 months to 7 years. Average follow-up 30 months

B. Indications for surgery

1. Abnormal head position (20° - 50°)
2. Abnormal head position and strabismus

C. Patients divided into 3 groups

1. Group 1 $n = 18$
 - Head turn without strabismus.
 - Bilateral surgery.
2. Group 2 $n = 7$
 - Head turn.
 - Strabismus.
 - Amblyopia.
 - Bilateral surgery.
3. Group 3 $n = 7$
 - Head turn.
 - Strabismus
 - Amblyopia
 - Unilateral surgery

D. Pre-Op evaluation

1. Estimate degree of head turn at distance while patient is binocularly viewing an accommodative target.
2. Visual acuity OD, OS, OU
3. Deviation (when present)
4. Versions
5. Worth 4-Dot
6. Titmus stereoacuity

E. Post-Op evaluation

1. Dosage of surgery

2. Degree of head turn
3. Visual acuity
4. Deviation
5. Versions
6. Worth 4-Dot
7. Stereoacuity

F. Results

1. Excellent = Residual head turn $\leq 5^\circ$
2. Good = Residual head turn $> 5^\circ$ but $\leq 15^\circ$
3. Poor = Residual head turn $> 15^\circ$

G. Dosage of surgery - Group 1

1. Parks' "Straight Flush" technique
 - ET eye - 5mm MR recession, 8 mm LR resection
 - XT eye - 7mm LR recession, 6 mm MR resection
2. Augmented technique (Calhoun & Harley - "Classic Plus")
 - a. Increase amount of surgery depending on degree of head turn.
 - b. Head turns 25° or less - 5, 6, 7, 8
 - $25^\circ - 45^\circ$ - 10-40% more surgery
 - 45° or more - 40%+ more surgery

H. Surgical results - Group 1

1. Classic - 9 patients
 - 67% = Good/Excellent
2. Augmented - 9 patients
 - 89% = Good/Excellent

I. Pre-op vs. post-op visual acuities - Group I

1. Improvement in binocular acuity 1-2 lines 11/18 (61%)
2. All patients had same or better postoperative stereoacuity

J. Dosage of surgery - Group 2.

1. Surgery for head turn
2. Amounts modified to correct strabismus at same time

K. Surgical results - Group 2

1. Augmented surgery 5/7
2. Good/Excellent post-op head position 3/7
3. Strabismus corrected $\pm 10^\Delta$ 6/7
4. Improved binocular visual acuity 3/7
5. Limited versions 6/7

L. Dosage of surgery - Group 3

1. Head turn and strabismus
2. Correction by doing R & R on one eye. i.e., R head turn, adducting eye fixing, esotropia present-R & R on fixing eye for both head turn and ET

M. Surgical results - Group 3

1. Good/Excellent post-op head positions 6/7
2. Strabismus corrected $\pm 10^\Delta$ 5/7
3. Limited versions 4/7

N. Summary

1. Head turns $\geq 25^\circ$ need augmented surgery
2. If strabismus is also present, plan to correct both
3. Some patients with head turn and strabismus can be corrected with unilateral surgery
4. Expect some improvement in visual acuity
5. Expect limitations of versions postoperatively when doing augmented surgery
6. Stability is based on limitations of versions - if present more stable

* Scott WE, Kraft SP. Surgical treatment of compensatory head position in congenital nystagmus. J Ped Ophthalmol Strab 1984;21(3):85-95.

Abnormal Head Positions Secondary to Motility Disorders

I. Abnormal Head Positions Secondary to Motility Disorders

A. Purpose of Adopting an Abnormal head Position

1. Fusion
2. Relief of diplopia
3. Obtain best visual acuity with nystagmus

B. Face Turn

1. VI nerve palsy
2. Duane's syndrome
3. Brown's syndrome
4. Inferior oblique palsy
5. Esotropia secondary to nystagmus
6. Periodic alternating nystagmus

C. Head Tilt

1. Superior oblique palsy-tilt to opposite side
2. DVD –tilt to same side
3. Congenital motor nystagmus

D. Chin Up/Chin Down

1. Monocular elevation deficiency
2. "A" and "V" patterns
3. Orbital floor fractures
 - a. Entrapment of inferior structures
 - b. Paresis of IR

4. Graves' ophthalmopathy
 - a. Inferior rectus restriction
 - b. Superior rectus restriction
 - c. Combination
5. Congenital fibrosis
6. Ptosis
7. Congenital motor nystagmus

E. Chin Down – Differential Diagnosis

1. "A" and "V" Patterns
 - a. "V" esotropia
 - b. "A" exotropia
2. Bilateral superior oblique palsy
 - a. If no secondary inferior oblique overaction - chin down.
 - b. If superior oblique palsy has inferior oblique overaction, there is usually no abnormal head position.
3. Orbital floor fracture - Inferior rectus paralysis

F. Chin Up - Differential Diagnosis

1. "A" and "V" patterns
 - a. "A" esotropia
 - b. "V" exotropia
2. Congenital fibrosis syndrome
3. Ptosis
 - a. An abnormal head position with ptosis is an indication for earlier surgery
4. Congenital motor nystagmus

5. Monocular elevation deficiency

6. Orbital Floor fractures

7. Graves' Ophthalmology

* Scott, WE, Weaver RG: Chin up and chin down head position. Amer Orthop J 1983;33:24-31

Supplement I

Amblyopia - Treatment & Results

Amblyopia - Treatment & Results

I. Definition: a difference in visual acuity of two or more lines between the eyes.

A. Functional amblyopia

1. Strabismic - secondary to disuse; most common
2. Deprivation - cataract, cloudy media; interferes with development of the fixation reflex
3. Ametropic - bilateral high refractive error
4. Anisometropic - a difference of greater than +1.00 D or -2.00 D

B. Organic amblyopia

A structural abnormality that is not treatable; i.e., optic nerve hypoplasia, posterior pole coloboma.

C. The incidence of unilateral amblyopia in children is 2-4%

McNeil (1955)	2.7%
Vereecken (1966)	4.4%
Gansner (1968)	2.39%
Kohler & Stigmar (1973)	2.2%

II. Visual Acuity Assessment in Children

A. Subjective tests

1. Snellen chart
2. Illiterate E
3. Stygar - no verbal response is needed, involves matching letters.
4. Allen Pre-School Pictures - may not always pick up a two line difference, 20/30 limit.
5. Isolated letters - amblyopes will show a better visual acuity secondary to the crowding phenomenon.

B. Objective tests

1. C S M Method - a difference in acuity is determined according to how well a patient maintains fixation on a particular target.

<u>Fixation Pattern</u>	<u>Visual Acuity</u>
Affixation	
Eccentric fixation	5/200 or less
Unsteady fixation	5/200 - 20/300
Central fixation but will not hold	20/200 - 20/100
Central fixation - will hold OU but prefers one eye	20/70 - 20/30
Alternates spontaneously	20/20 OU

Cross fixation - does not necessarily imply equal visual acuity.

- a cross-fixator must switch at the midline or amblyopia should be suspected.

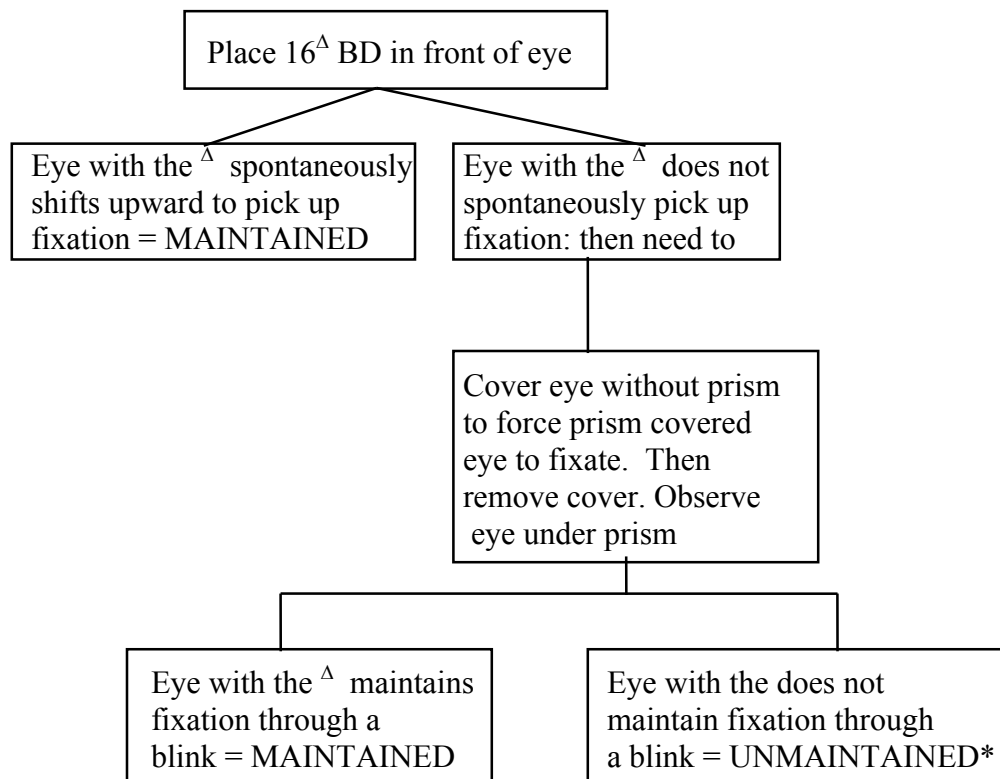
2. Binocular fixation pattern method

Use cover/uncover test - observe uncovered eye.

- If the uncovered eye fixates and holds when the covered eye is uncovered for a measured time but not to a blink - visual acuity is usually within one line of the other eye.
- If the uncovered eye holds fixation to a blink - there is usually less than one line difference.
- If the uncovered eye holds fixation through a blink - visual acuity is equal.
- If the uncovered eye deviates immediately upon removal of cover - there is usually a 2 or more line difference and amblyopia should be treated.

* Zipf RF. Binocular fixation pattern. Arch Ophthalmol 1976;94:401-405.

- 16^Δ prism induced deviation - base down or base in, if base down can't be done - used in patients who don't have manifest strabismus.



If patient holds

- briefly → a significant amblyopia is present of more than two lines.
- to but not through a blink → mild amblyopia of two lines exists.
- not at all dense → amblyopia.

The prism is then placed over the opposite eye and the same steps are followed. If the response to this test is the same with each eye, both eyes are considered to have equal visual acuity.

* Wright KW, Walonker F, Edelman P. 10-Diopter fixation test for amblyopia. Arch Ophthalmol 1981;99:1242-1246.

4. Forced preferential looking: This test consists of a series of cards, which contain a pattern stimulus and a homogeneous, non-pattern stimulus. The cards are held at a given distance from the subject. The examiner, who cannot see which side of the card is the patterned side, observes the direction that the baby looks and records this.

If the stimulus is large enough to be visible to him/her, a young baby will look at the pattern rather than the homogeneous stimulus. The stimuli are presented in decreasing size until no preference is elicited by the baby. At this stage, it is assumed that the baby can no longer see the stimulus and the level is recorded.

* When a child becomes distracted by background objects, noises, etc., they are becoming too old for the test.

III. Amblyopia Diagnosis

- A. Observe more than once during exam
- B. Fixation pattern is dependent on target size
- C. Always check near vision
- D. If patient comes in wearing patch check fixation at least 30 minutes after patch has been removed
- E. Small angle ET's (monofixators) usually have a fixation preference without significant amblyopia
- F. Cross-fixators should switch at midline
- G. Latent nystagmus is not a contraindication for amblyopia treatment
- H. Amblyopia is uncommon in exo and vertical deviations.

IV. Occlusion

- A. Full-time occlusion (FTO) = patching 24 hours/day
- B. Little risk of occlusion amblyopia, but if it does occur, it is easily reversed
- C. Follow-up at intervals of 1 week per year of age of the child
- D. End-point: visual acuity is equal or 3 consecutive episodes of compliant FTO render no improvement
- E. FTO should be followed by part-time occlusion (PTO) 6-8 hours/day
- F. PTO may be tapered and discontinued when visual acuity remains stable without patching, with follow-up every 3-4 months
- G. Slippers should be kept on PTO until visual maturity

V. Results of strabismic amblyopia therapy

- A. Age of patient at the time treatment is started is usually related to duration of treatment; i.e., the older a patient, the longer it will take to achieve equal visual acuity
- B. The age of the patient at the time treatment is started usually does not affect the final visual result

C. Of 78 patients in various age groups 49 did not slip after patching was discontinued

D. Patients in whom equal vision is obtained before age six are no more likely to slip when patching is discontinued than those achieving equal visual acuity after age six

E. Final visual acuity generally is not influenced by initial visual acuity. *Exception if initial 20/100 or worse, less likely to have final visual acuity of 20/20.

F. Results of full time occlusion

Excellent (20/20)	76%
Good (20/25 - 20/40)	16%
Fair (20/50 - 20/100)	5%
Poor (<20/100)	3%

G. Complications of patching

1. Social pressure.
2. Skin irritation.
3. Occlusion amblyopia.

* Scott WE, Dickey CF. Stability of visual acuity in amblyopic patients after visual maturity. Graefes Archiv Clin Exp Ophthalmol 1988;226:154-157.

* Scott WE, Stratton VB, Fabre J. Full-time occlusion therapy for amblyopia. Amer Orthop J 1980;30:125-130.

VI. Results of anisometropic amblyopia therapy

A. Definition - anisometropia

1. Hyperopia (H) - $\geq +1.00$ D
2. Myopia (M) - ≥ -2.00 D
3. Astigmatism (A) - $\geq +1.00$ cyl.
4. Compound (C) - $\geq +0.75$ cyl. & sph.
 - a. Hyperopic Astigmatism (CHA)
 - b. Myopic Astigmatism (CMA)

B. Patient population

<u>Type</u>	<u>n</u>	<u>strab</u>	<u>non-strab</u>
M	23 (19%)	13	10
H	42 (34%)	9	33
A25 (20%)	12	13	
CMA	18 (14%)	9	9
CHA	16 (13%)	6	10
Total	124	49 (40%)	75 (60%)

C. Hyperopic differences in refractive error (mean +2.46 D) more amblyogenic than myopic differences (mean -8.05 D)

D. The results of treatment were influenced by

1. Initial visual acuity.
2. Type of anisometropia - myopes and CMA had poorer visual outcomes.

E. The final visual acuity was not influenced by

1. Patient age at the time of treatment onset.
2. Presence or absence of strabismus.
3. Type of initial treatment - glasses with or without patching.

F. Overall, 83% obtained 20/40 or better visual acuity

* Kutschke PJ, Scott WE, Keech RV. Anisometropic amblyopia. Ophthalmol 1991;98(2):258-263.

VII. Results of deprivation amblyopia therapy

A. Visual acuity of monocular cataracts and persistent hyperplastic primary vitreous (PHPV)

VA Results

	Excellent >20/50	Good 20/60-20/100	Poor <20/100
%	43	29	29
Ave. Age of Surgery (Range)	5.4 weeks (9 days- 14 weeks)	8.8 weeks (5weeks-15 weeks)	20 weeks (7 weeks, 9 months)
Ave. Age of Optic Correction (Range)	7 weeks (15 days- 17 weeks)	17 weeks (7weeks-28 weeks)	24 weeks (7weeks-9 months)
Patching Compliance	Excellent	Excellent-Poor	Poor
Contact Lens Wear Compliance	Excellent	Excellent-Poor	Poor

B. Patching program

First 2mos of life	50% PTO
2mos - 7mos	75% PTO
7mos - Subjective V/A	100% FTO
Stable V/A-Visual Maturity	50% PTO/FTO

Monitor:

Binocular Fixation Pattern
Reverse Patch 50% for Fixation Switch
Resume PTO/FTO when phakic eye preferred

C. Successful visual development depends on

1. Surgical removal of PHPV or MCC early (before 3 months of age)
2. Placement of optical correction (high plus powered contact lens)
3. Early onset of occlusion therapy with good compliance.

* Karr DJ, Scott WE. Visual acuity results following treatment of persistent hyperplastic primary vitreous. Arch Ophthalmol 1986;104(5):662-667.

* Drummond GT, Scott WE, Keech RV. Management of monocular congenital cataracts. Arch Ophthalmol 1989;107(1):45-51.

VIII. Results of Organic Amblyopia Therapy

A. 51 patients studied

1. 14 - optic nerve anomalies.
2. 25 - media opacities.
3. 12 - macular lesions.

B. Results

1. Of all patients, 39% had visual acuity 20/40 or better following treatment.
2. Earlier age at presentation associated with better results in patients with media opacities and macular lesions, but not in patients with optic nerve abnormalities.
3. No significant differences found between those whose vision increased to better than 20/200 and those whose vision did not increase in the areas of:
 - a. Presence or absence of strabismus.
 - b. Presence or absence of anisometropia.
 - c. Presence or absence of RAPD (ON group only).
4. No patients with posterior lenticonus improved vision with treatment.
5. No patient with optic nerve hypoplasia increased visual acuity to better than 20/200 with treatment.

*Bradford GM, Kutschke PJ, Scott WE. Results of amblyopia therapy in eyes with unilateral structural anomalies. *Ophthalmol* 1992;99(10):1616-1621.