Pediatric Strabismus

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How big of a problem is strabismus?
Population Based Studies

- Largely occur through school based studies-
- may underestimate developmentally delayed children or those attending a private school
- Clinic based studies have a referral bias
- Preschool studies are relatively uncommon due to difficulty identifying schools
Population Based Studies of Preschool Age Children

- **Baltimore Pediatric Eye Disease Study**
  - Population based study of preschool aged White and African American in the Baltimore, MD area revealed that manifest strabismus affected 1 in 30 white (3.3%) and 1 in 47 (2.1%) African American children in this age group.

- **The Multi-ethnic Pediatric Eye Disease Study**
  - Population based study of preschool aged Hispanic and African American children in the Los Angeles area reported the prevalence of strabismus of 2.5% in both groups.
Population Based Studies

Conclusions from both studies

National population projections suggest approximately 677,000 cases of manifest strabismus in children 6-71 months
Olmstead County, Minnesota, 1994

• Retrospective Chart Review of all patients younger than 19 years of age diagnosed with esotropia from 1985-1994

• Annual incidence identified as 111.0 cases of childhood esotropia per 100,000 patients younger than 19 years of age

• approximately 2.0% prevalence of esotropia in patients 5 years old and younger in 10 year time period
Why is it important to identify and treat strabismus early?

• prevent the development of amblyopia
• restore binocularity - aid in the development of stereopsis
• improve treatment outcomes
What are the risks of uncorrected strabismus?

- Amblyopia
  - Increased rate of severe bilateral vision loss as patients with amblyopia are more likely to suffer injury to the healthy eye
- Decreased Stereovision
  - Disqualification for applicants of Class I Air force Pilot and Naval Aviator
  - Qualifications for surgical subspeciality training?
- Psychosocial Discrimination
Risks of uncorrected strabismus

Psychosocial Aspects of Strabismus Study

Denise Satterfield, MD; John L. Keltner, MD; Thomas L. Morrison, PhD

The psychosocial aspects of strabismus in teenagers and adults and the impact of surgical correction

Bradley A. Nelson, Kammi B. Gunton, MD, Judith N. Lasker, PhD, Leonard B. Nelson, MD, and Lea Ann Drohan, MD
• Esotropia
  • Infantile/Congenital
  • Duane Syndrome
  • Moebius Syndrome
  • Accomodative
  • Nystagmus Blockage Syndrome
  • CNVI Palsy

• Exotropia
  • Infantile
  • Intermittent
  • Duane Syndrome
  • CNIII Palsy

• Hypertropia
• CNIV Palsy
• Hypotropia
• Entrapped IR
• Double Elevator Palsy
• Syndromes
• Congenital Fibrosis Syndrome
• CPEO
• Craniosynostosis
• Myasthenia Gravis
Esodeviations

nonfixating eye deviates in
Congenital Esotropia

- Present by 6 months of age
- Distance and near measurements equal
- Normative refractive error for age
- Large angle of deviation, often alternating
- Amblyopia uncommon
- Associated with DVD and IOOA - present by 3-4 years of age
- Family history common
Right Inferior Oblique Overaction
DVD

- Slow upward drifting of the nonfixating eye
- Constant in all positions of gaze - no pattern
- Movement is upward, abduction and excyclotorsion
- Can be unilateral, bilateral, assymetric
- No compensatory hypodeviation in the contralateral eye (violates Herring’s Law)
Accommodative Esotropia

- Develops between 6 months and 7 years
- Can appear acutely, or start out intermittent
- Associated with high hyperopic refractive error
  - $+3.00-+10.00$ D (average $+4.75$ D)
- High incidence of amblyopia
- Distance near deviation similar
High AC/A Ratio

- ET near > distance (difference > 10 D)
Mixed Mechanism and Nonaccomodative Esotropia

- **Partially Accomodative**
  - esotropia only partially controlled by full hyperopic correction
  - residual esotropia treated with eye muscle surgery

- **Nonaccomodative**
  - acquired, occurring in older children
  - unknown etiology, nml refractive error
  - most rule out associated neurologic disease
Duane’s Syndrome

Failure of formation of the abducens nucleus and sixth nerve, innervation of the lateral rectus by an anomalous branch of the third nerve within the orbit
• Female > Male

• OS > OD > OU

• ET > Ortho > XT

• A-V patterns common

• Anisometropic Amblyopia

• Associations: **Goldenhaar Syndrome** (limbal dermoid/lipodermoid, preauricular skin tag, superior eyelid coloboma), **deafness**, **crocodile tears** (nerve fibers from mandibular and sublingual gland reinnervate the lacrimal gland)
Congenital or Acquired Cranial Nerve VI Palsy

- **Congenital**
  - rare, well documented
  - prognosis for recovery good
  - focal damage to the peripheral nerve, but nerve still intact

- **Acquired**
  - etiologies include: traumatic, increased ICP, meningitis
Acquired CN VI Palsy

Right CN VI Palsy
With optic nerve edema
Differentiating CN VI Palsy from Duane Syndrome

Right CN IV Palsy

Left Type 1 Duane Syndrome with Eyelid fissure narrowing in adduction
Less Common Esodeviations

- Nystagmus Blockage Syndrome
  - convergence dampens nystagmus, pts fixate with adducted eye, causing head turn to side of deviating eye

- Mobius Syndrome
  - CN 6, 7, 9 palsy
  - “mask like facies”
  - limb, chest and tongue defects
  - ET or ortho
Exodeviations

nonfixating eye deviates out
Types of Exodeviations

- **Congenital Exotropia**
  - Occurs under the age of 1 year - rare
  - Angle of deviation is large, average 35 prism diopters
  - Amblyopia is not common
  - Similar refractive error to general population
  - Early surgical correction indicated
Types of Exodeviations

• **Sensory exotropia**
  - Secondary deviation due to poor vision in one eye
    - Uncorrected refractive error, amblyopia, media opacity, organic lesion

• **Intermittent Exotropia**
  - Onset varies: infancy to 4 years of age
  - Most common form of exotropia
    - Outward drifting of one eye, interspersed with periods of good alignment
  - May be progressive - may start out as phoria, progress to intermittent exotropia, and then become manifest exotropia
Prior to cover testing

After cover testing
Cranial Nerve III Palsy - congenital and acquired

• **Congenital**
  • rare, most commonly unilateral
  • ptosis, exotropia, hypotropia
  • **smaller pupil on affected eye**
  • midbrain maldevelopment, intrauterine injury, perinatal damage
  • 50% have associated neurologic signs

• **Acquired**
  • trauma, infection, tumor and vascular etiologies
  • damage of nerve as it courses subarachnoid space
Hypertropias
(nonfixating eye elevates)
Congenital or Acquired Cranial Nerve IV Palsy

- Most common congenital oculomotor palsy
- Half of all isolated CN IV palsies are congenital
- Most common presentation is with abnormal head tilt
- Vertical deviations usually large (av. 18-20 pd)
- Large vertical fusional amplitudes, up to 20 D
- Hypodevelopment of contralateral face
- May decompensate later in life
Congenital CN IV Palsy

- **Etiology:**
  - not associated with prenatal disorders or birth trauma
  - possible underdevelopment of the superior oblique tendon, or muscle
Hypertropia increases in ipsilateral head tilt and contralateral gaze.
Hypotropia

nonfixating eye deviates downward
Brown Syndrome

- Limitation of elevation in adduction
- Relatively normal elevation in abduction
- V pattern exotropia in upgaze
- No overacting SO (distinguishes from IO palsy)
- Etiology: restriction of the SO tendon, or the trochlea/tendon complex
- Congenital > Acquired
  - Acquired: SO tuck, valve or buckle near SO, inflammatory, infectious, traumatic in the trochlear region
No elevation of the adducted right eye
Monocular Elevation Deficit
(formerly known as Double Elevator Palsy)

Paresis of both inferior oblique and superior rectus
Monocular Elevation Deficit

- Hypotropia, limitation of elevation abduction and primary upgaze
- Ptosis (in 50% of pts) and pseudo-ptosis present
- Chin up head posturing
KNAPP Procedure
Transposition of rectus muscles

- If IR tight, recess IR
- If IR not tight, transpose MR and LR to SR (re-create supraduction vector) - KNAPP
- Can be half tendon width or full tendon
Orbital floor fracture with entrapped inferior rectus

- Inferior Rectus becomes incarcerated in the fracture, or tethered on a bony fragment and restricted in a hypotropiopic position
- Limitation in upgaze, + forced duction test
Strabismus Syndromes
**Congenital Fibrosis Syndrome**

- Group of congenital anomalies with variable restriction of EOM
  - + FDT due to restriction
- Nonprogressive
  - Congenital fibrosis - most severe, AD
  - congenital fibrosis of inferior rectus
  - Strabismus fixus
  - Vertical retraction syndrome
  - Congenital unilateral fibrosis
Chronic Progressive External Ophthalmoplegia (CPEO)

- sporadic, or mitochondrial
- may present at any age
- severe ptosis with complete ophthalmoplegia, no restriction on FDT, no Bell’s (because not supranuclear)
- association: Kearns-Sayre Syndrome
Congenital Myasthenia Gravis

- Presentation from birth to early adulthood - most commonly in first 2 years
- Sporadic or AD inheritance, not autoimmune
- Presentation of bilateral ptosis, facial weakness, variable limitation in eye movements, possible limb weakness, and reduced muscle mass
- May show some response to anti-cholinesterase tx
Juvenile Autoimmune Myasthenia Gravis

- Age of onset < 15 years
- Female to male ratio - 3:1
- Presentation with ocular MG, 50% progress to generalized MG
- at greater risk than adult disease for aspiration and ventilator dependency
- Associations: Hyperthyroidism, Juvenile onset DM, Rheumatoid Arthritis
- Treatment with anti-cholinesterase drugs, immunosuppression, plasmapheresis
Craniosynostosis

- Premature closure of a cranial suture which results in deformities of skull shape
- Orbital and ocular extorsion,
  - Causes varying patterns of strabismus
    - MR – adducts and elevates (simulates IOOA, SOUA)
    - SR – elevates and abducts
    - LR – abducts and depresses
    - IR – depresses and adducts
Crouzon Syndrome

Apert Syndrome
Excision of Muscle

Marking of inferior and superior pole, checking of excised muscle
Rectus Muscle Recession

Muscle belly is pulled up to position at its new insertion site
Untreated or recurrent childhood strabismus can lead to strabismus in adulthood
Adult vs. Pediatric Strabismus

• Onset before visual maturation
  • Adapt to misalignment with SUPPRESSION or abnormal retinal correspondence (ARC)
  • No diplopia!
  • Untreated or recurrent childhood strabismus makes up majority of adult strabismus patients

• Onset after visual maturation
  • Onset after age 8-10 years
  • Cranial nerve palsy, thyroid related eye disease, traumatic
  • Misalignment causes diplopia, patients may adopt abnormal head posture
Why treat adult strabismus?

• Resolution of diplopia
• Improved depth perception
• Expand peripheral field of vision
• Psychosocial aspects of reconstructed ocular alignment
• Emotional, social and economic benefits
How prevalent is adult strabismus?
2012 review of random Medicare Part B physician claims from 2002-2010 performed to determine the prevalence of strabismus and strabismus surgery in the aged medicare fee-for service population.

0.68 % of aged Medicare beneficiaries were diagnosed with strabismus in 2010. Strabismus surgery was performed on 0.016% of beneficiaries, or in 2.3% of those patients diagnosed with strabismus.

Why are so few patients undergoing strabismus surgery?
Why delay surgical correction?

27 % surgery was never offered
23% surgery was offered, but patient declined
13% prior satisfaction with nonsurgical care
11% care never sought after
6 % prior poor surgical experiences
6% told by eye care specialist that nothing could be done, or surgery could make things worse


**Reasons for delay of surgical intervention in adult strabismus.**

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