EXOTROPIA

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DIAGNOSING EXOTROPIA

The first step in safely and successfully looking after patients presenting to you with XT is to differentiate[1]:

- Those caused by potentially dangerous ocular, visual or neuronal pathology (up to 33%)

from

- Those that are primary, benign exotropia (67% or more)

Note that it is likely optometry sees a far higher proportion of primary, benign exotropias than this study tertiary care ophthalmology population.

[1]Data from tertiary care USA – Mohney & Huffaker in Ophthalmol 2003; Govindan, Mohney et al in Ophthalmol 2005

NEUROLOGICAL AND PATHOLOGICAL EXOTROPIAS

In a tertiary care ophthalmology setting in the USA, Mohney and Huffaker (2003) found:

- 21% of first presenting exotropia was associated with CNS abnormalities, some of which were long-standing (e.g. cerebral palsy, developmental delay, Down Syndrome) and some of which were new/emergent and potentially dangerous (e.g. hydrocephalus, cerebral haemorrhage, motor vehicle accident, meningitis)
- 10% had sensory exotropia (lack of fusion due to monocular vision loss, leading to exotropia)
- 2% had paralytic exotropia (either Duane's Syndrome Type 2, or third cranial nerve palsy)

Figure 14.1: Complete CN3 palsy leaves an eye pointing "down and out" because only the LR and SO muscles work (unopposed). There will also be a ptosis and a pupil defect.

Differentiating these potentially dangerous exotropias for referral requires describing direction, frequency, magnitude, laterality, comitancy of direction and comitancy of distance, together with (at minimum):

i. Taking a relevant history that establishes the nature of onset and searches for any relevant associations
   - Unusual symptoms that will help you to differentiate out potentially dangerous XTs (and refer them appropriately): Nausea and/or vomiting, unusual headaches, clumsiness, general malaise, trauma, recent weight changes, hormonal changes, growth defects

ii. Pupil check – both afferent and efferent pupil pathways
   - e.g. An optic nerve glioma could cause an exotropia and causes an afferent pupil defect; a CN3 palsy causes an exotropia and can cause an efferent pupil defect) – refer appropriately

iii. Observation of the external eye with specific scrutiny for any proptosis
   - e.g. Rhabdomyosarcoma can cause XT via proptosis and requires urgent referral

iv. Ocular health examination (eye disease can cause sensory exotropia, and may need urgent referral)
   - Best-practice ocular fundoscopy – preferably through dilated pupils – is essential
   - Checking the clarity of the ocular media

v. Checking for any other unusual signs suggestive of neurological or pathological causes of XT
   - E.g. Nystagmus, torticollis, ptosis – refer appropriately

PRIMARY, BENIGN EXOTROPIAS

In a tertiary care ophthalmology setting in the USA, Mohney and Huffaker (2003) found:

- 3% had infantile exotropia, half of which spontaneously resolved by 4 months of age
- 4% were unclassifiable, but had a high incidence of amblyopia and/or cyclo-vertical disturbances, and were usually small angle strabismus
- 12% had convergence insufficiency (CI = larger exo-deviation at near than distance)
- 48% had intermittent exotropia (could be CI, basic (same deviation at distance and near), or divergence excess (DE = larger exo-deviation at distance than near))

The aetiology of primary, benign exotropia is not as clearly defined as that of esotropia. Some authors (e.g. Duane) have suggested the cause is bad innervation (either lack of convergence drive, or too much active divergence). Other authors (e.g. Bielschowsky) have suggested the cause is imbalanced mechanical forces (either orbital shape, tendon length, attachment position, or a combination of these).

- No definitive cause has emerged, despite the efforts of authors such as Tamler and Jampolsky (1967) and Crosby (1953)
- It seems likely that innervational and mechanical factors are probably both important to different degrees in different exo-deviations

More importantly to the clinician, aetiology is not as important in deciding management for primary, benign exotropias as it is for esotropias. In exotropia, clinical decision-making will be assisted by the following:

i. Differentiating whether the exotropia is CI, basic, or DE?
   - Measure the size of the XT at distance and near
   - Different treatments are appropriate depending on distance/near incomitancy

ii. Differentiating whether the exotropia is constant or intermittent?
   - Determine constancy with unilateral cover test
   - Different treatments are appropriate for intermittent versus constant exotropia
   - Prognosis is better for intermittent exotropia

iii. Differentiating whether the exotropia is unilateral or alternating?
   - Determine laterality with the unilateral cover test
   - Alternating exotropia means there will be no or minimal amblyopia, which means different treatments will be appropriate

iv. Differentiating whether the exotropia has any vertical deviation?
   - Use the alternating cover test to look for any vertical deviation (in any direction of gaze, but particularly primary gaze)
   - Vertical deviations make it very difficult to achieve fusion – the vertical deviation may need to be corrected (with prism or surgery) before other treatments for exotropia can work

v. Careful refraction

- Clear vision that is as equal as possible between right and left eyes assists fusion. Therefore careful refraction (this may mean non-cycloplegic retinoscopy, cycloplegic retinoscopy, subjective refraction, blur function refraction, etc) is critical

vi. Quantification of amblyopia, including measurement of sensory fusion capacity

- Test visual acuity (single symbol and logMAR)
- Test for fusion/suppression/diplopia/anomalous correspondence using Worth 4 Dot, and/or stereotest suppresion check
- Test for high grade fusion using contour stereoacuity (e.g. using the Randot circles) and random dot stereopsis (e.g. using the Randot shapes)

vii. Measure vergence ranges (in any patient with intermittent exotropia)

- From the fused state, how much prism challenge can you give before blur/break/recovery occur (see accommodation vergence chapter for details, but beware you may need some adjustments in patients with intermittent strabismus)
- Compare the vergence range with the deviation to see if Sheard's Criterion is satisfied or not

MANAGING PRIMARY BENIGN EXOTROPIA

When you have…

- Differentiated between CI XT, basic XT and DE XT
- Differentiated intermittent from constant, unilateral from alternating, & comitant from incomitant XT
- Determined the magnitude of the deviation
- Quantified amblyopia, fusion, vertical deviations

…then you are in a position to consider the management options for primary, benign exotropia. Success of each option changes with these variables. The management options are: vision therapy, minus lens over-correction, alternate occlusion, prism therapy, and surgery.

OPTION 1 – VISION THERAPY

Vision therapy aims to decrease suppression, improve fusion per se, and in particular improve fusional convergence. Techniques include red/green anti-suppression activities, smooth and jump vergences with Brock String or equivalent, loose prism activities, tranaglyph targets, aperture rule work, and/or glasses with plus lenses and BO prism.

Coffey et al's (1992) review paper estimated the success rate for treating exotropia with vision therapy to be 59%.Vision therapy is considered more successful when XT is:

- Intermittent rather than constant
- CI rather than DE

Sheiman et al (2005 and 2008) performed randomised clinical trials of CI treatments in children:

- An intense vision therapy program that challenges fusional vergence (using either loose prisms, computer-based disparity, aperture rule or tranaglyphs) while holding viewing distance (i.e. proximal and accommodative drives to vergence) steady, improves signs and symptoms of CI

- A less intense pencil push-up program (such as that suggested by many non-specialist optometrists and ophthalmologists) does not improve signs or symptoms of CI

- An intense office-based placebo program did not improve signs or symptoms of CI

** These studies changed ophthalmology's view of optometric vision therapy – as described by Burt Kushner in Archives of Ophthalmology 2005 **

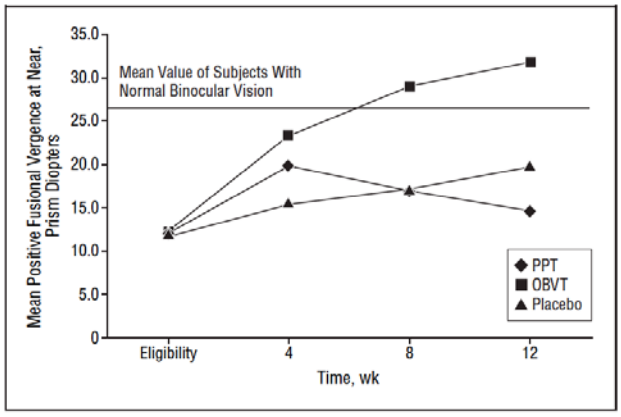

Figure 6. Mean positive fusional vergence at near after 4, 8, and 12 weeks of treatment. PPT indicates pencil push-ups therapy; OBVT, office-based vision therapy.

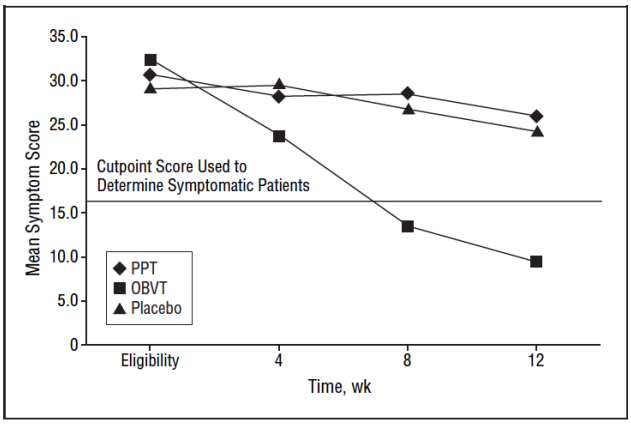

Figure 4. Mean convergence insufficiency symptom score after 4, 8, and 12 weeks of treatment. PPT indicates pencil push-ups therapy; OBVT, office-based vision therapy.

Figure 14.2 : Improvements in mean positive fusional vergence and symptoms, per week, according to different treatment options

A two stage guide to vision therapy for exotropia:

Stage 1 – Overcome suppression

- Use Monocular Fixation in a Binocular Field (MFBF), in which red/green filters and targets allow the patient to see the whole room with both eyes, but provide specific visual detail of interest to only one eye (the eye that usually suppresses)

- Use Brock String with or without red-green filters to use physiological diplopia to overcome suppression

Stage 2 – Improve fusion

- Note Horwood and Torr's (2014) finding that it is better to work on vergence and accommodation seperately –i.e. loose prism therapy, tranaglyphs, computer-based therapy, aperture rule work, loose lenses and flipper work are all likely to be more successful than pencil pushups or Brock String (in Stage 2)

- Aim to increase fusional convergence (BO reserves) until Sheard's Criterion is satisfied

OPTION 2 – MINUS LENS OVER-CORRECTION

By prescribing less plus or more minus than your refraction suggests, minus lens over-correction forces a patient to accommodate in order to make vision clear. Accommodative convergence is then also stimulated, thereby decreasing the exo-deviation (by an amount predictable from AC/A ratio), and hopefully providing an opportunity for fusion to develop.

- Caltrider and Jampolsky (1983) had some level of success in 72% of intermittent exotropes they treated with minus lens over-correction
- Many practitioners worry about inducing myopia (which is a reasonable concern from animal models, although there is no human data suggesting this will happen)
- Many practitioners worry that minus lens over-correction may cause more discomfort in the long-term rather than less
- It can be considered justifiable if:
    - A patient has a high AC/A ratio (i.e. there is good (vergence) reward for (accommodative) effort), AND you use it in conjunction with other treatments (i.e. you don't intend to use it forever)

OPTION 3 – ALTERNATE OCCLUSION

Part-time alternating occlusion is said to improve control of the deviation by working against suppression mechanisms. Improved control seems to be temporary (Flynn, McKenney and Rosenhouse, 1975), so this is not a recommended treatment.

OPTION 4 – PRISM THERAPY

Use Sheard's Criterion to determine the amount of prism you need to prescribe in spectacles to achieve clear, comfortable, single binocular vision. Coffey et al's (1992) review paper estimated a success rate of 28%. Prism therapy works best for adults with intermittent XT who experience diplopia when tropic (no suppression suggests good fusion capacity; and the deviation is only sometimes too big for fusional convergence skills).

OPTION 5 – SURGERY

Surgery aims to decrease the magnitude of the exo-deviation by some combination of LR recession & MR resection.

- Optimal stereoacuity prior to surgery promotes long-term stability of alignment – i.e. there is a role for vision therapy combined with surgery (Birch et al 2004)
- Surgery is most useful for large angle XT (as non-surgical approaches become ineffective)
- Surgery appears to be more effective with emmetropic and hyperopic patients than with myopic patients (Gezer et al in JAAPOS 2004)
- Coffey et al's (1992) review paper estimated a success rate of 46% for surgery
- A recent Greek study from a single institution reported a success rate (from a single surgery) of 69% (Aslanis et al, Fr J Ophthalmol 2006)
- Abroms, Mohney, Parks et al (Am JOphthalmol 2001) found that time of surgery impacts the outcome success – so don't delay surgical opinion indefinitely.

REFERENCES

EXOTROPIA REFERENCES

McKean-Cowdin R, et al. Prevalence of amblyopia or strabismus in asian and non-Hispanic white preschool children: multi-ethnic pediatric eye disease study. Ophthalmol 2013;120(10):2117-24

Multi-ethnic Pediatric Eye Disease Study Group. Prevalence of amblyopia and strabismus in African American and Hispanic children ages 6 to 72 months the multi-ethnic pediatric eye disease study. Ophthalmol 2008;115(7):1229-36

Donnelly UM. Horizontal strabismus worldwide--what are the risk factors? Ophthalmic Epi 2012;19(3):117-9

Yu CB, Fan DS, Wong VW, Wong CY, Lam DS. Changing patterns of strabismus: a decade of experience in Hong Kong. British J Ophthalmol 2002;86(8):854-6

Ntim-Amponsah CT, Ofosu-Amaah S. Prevalence of refractive error and other eye diseases in schoolchildren in the Greater Accra region of Ghana. J Ped Ophthal Strab 2007;44(5):294-7

Mohney and Huffaker. Common forms of childhood exotropia. Ophthalmol 2003;110:2093-2096.

Govindan, Mohney, Diehl and Burke. Incidence and types of childhood exotropia. Ophthalmol 2005;112:104-108.

Tamler and Jampolsky. Is divergence active? An electromyographic study. Am J Ophthalmol 1967;63:452.

Crosby. Relations of brain centres to normal and abnormal age movements in the horizontal plane. J Comp Neurol 1953;99:437

Coffey, Wick, Cotter, Scharre and Horner. Treatment options in intermittent exotropia: a critical appraisal. Optom Vis Sci 1992;69(5):386-404

Scheiman M, CITT group et al. A survey of treatment modalities for convergence insufficiency. Optom Vis Sci 2002;79(3):151-7

Horwood A, Toor S. Clinical test responses to different orthoptic exercise regimes in typical young adults. Ophthalmic & Physiological Optics 2014;34(2):250-62

Scheiman M, CITT group et al. A randomised clinical trial of treatments for CI in children. Arch Ophthalmol 2005;123:14-24

Kushner BJ. The treatment of convergence insufficiency. Arch Ophthalmol 2005;123(1):100-1

Caltrider and Jampolsky. Overcorrecting minus lens therapy for treatment of intermittent exotropia. Ophthalmol 1983;90:1160-1165

Flynn, McKenney, Rosenhouse. A method of feating intermittent divergence strabismus. In Gnanaraj and Richardson. Interventions for intermittent distance exotropia: review. Eye 2004:1-5

Sheard C. Zones of ocular comfort. Am J Optom 1930;7:9-25

Birch, Stager, Berry and Leffler. Stereopsis and long-term stability of alignment in esotropia. Journal of AAPOS 2004;8:146-150

Gezer, Sezen, Nasri, Gözüm. Factors influencing the outcome of strabismus surgery in patients with exotropia. Journal of AAPOS 2004;8:56-60

Aslanis D, et al. [Surgical results in childhood primary comitant large-angle exotropia]. Journal Francais d'Ophtalmologie. 2006;29(1):37-42

Abroms, Mohney, Rush, Parks and Tong. Timely surgery in intermittent and constant exotropia for superior sensory outcome. Am J Ophthalmol 2001;131:111-6