American Orthoptic Journal

ISSN: 0065-955X (Print) 1553-4448 (Online) Journal homepage: https://www.tandfonline.com/loi/uaoj20

Beware: Armed and Dangerous—Acquired Non-Accommodative Esotropia

Heather Macpherson, Inge De Becker & James R. MacNeill

To cite this article: Heather Macpherson, Inge De Becker & James R. MacNeill (1996) Beware: Armed and Dangerous—Acquired Non-Accommodative Esotropia, American Orthoptic Journal, 46:1, 44-56, DOI: 10.1080/0065955X.1996.11982070

To link to this article: https://doi.org/10.1080/0065955X.1996.11982070

Published online: 05 Apr 2018.

Beware: Armed and Dangerous—Acquired Non-Accommodative Esotropia

Heather Macpherson, O.C.(C.), COMT[1]
Inge De Becker, M.D., FRCS(C)[1,2]
James R. MacNeill, M.D., FRCS(C)[1,2]

ABSTRACT

Acquired non-accommodative esotropia is generally considered to be a benign phenomenon. However, numerous instances of posterior fossa tumors initially presenting as acute comitant esotropia have been reported. The presence of any of the following warrant further investigation when associated with acute onset, non-accommodative esotropia: nystagmus, vertical strabismus, other cranial nerve involvement, parental concerns of poor motor control, headache or persistent diplopia.

Other types of acquired non-accommodative esotropia include decompensated esophoria/monofixation syndrome, VI N paresis, divergence paresis, myasthenia gravis and mechanical/restrictive strabismus resulting from Graves disease, trauma, iatrogenesis and orbital myositis are discussed. Clinical investigation and treatment are briefly reviewed.

From the Department of Ophthalmology, IWK-Grace Health Centre[1] and the Department of Ophthalmology, Dalhousie University,[2] Halifax, Nova Scotia, Canada.

Request for reprints should be addressed to: Heather Macpherson, 6S, IWK-Grace Health Centre, 5850 University Ave., Halifax, Nova Scotia, Canada, B3J 3G9.

Presented as part of a Symposium at the Joint Meeting of the American Orthoptic Council, the American Association of Certified Orthoptists and the American Academy of Ophthalmology Atlanta, Georgia, October 29, 1995.

INTRODUCTION

It is certainly not uncommon for patients to present to a strabismus clinic with acquired non-accommodative esotropia. The aim of this paper is to help the clinician differentiate the benign from the ominous pathologic. The clinical investigation of patients with acquired esotropia will be discussed with some treatment options briefly introduced. Table 1 outlines differential

TABLE 1
DIFFERENTIAL DIAGNOSES OF ACQUIRED NON-ACCOMMODATIVE ESOTROPIA

- Secondary to Vision Loss/Sensory ET
- Decompensated esophoria/monofixation syndrome
  —following occlusion or other disruption of binocular vision
  —spontaneous decompensation (Burian—Franceschetti type)
- Neurogenic
  —abducens paresis
  —intracranial disease
  —divergence paresis
  —myasthenia gravis
- Mechanical
  —Graves (endocrine) ophthalmopathy
  —orbital wall fracture
  —iatrogenic (i.e. post surgery)
  —orbital myositis
- Refractive
  —progressive myopia
  —overcorrected myopia
- Other
  —cyclic esotropia
  —voluntary esotropia/spasm of the near reflex

diagnoses of acquired non-accommodative esotropia.

ESOTROPIA SECONDARY TO VISION LOSS/SENSORY ESOTROPIA

Significantly decreased visual acuity resulting from anisometropia or any pathologic condition in one eye poses an obstacle to fusion, and will often, but not always, lead to a sensory strabismus. Most commonly seen is sensory strabismus resulting from ocular injury, corneal opacity, unilateral cataract, macular lesion or optic atrophy.[1] In our hospital three of the last five patients diagnosed with retinoblastoma had esotropia as an initial sign, though two of those patients later developed leukocoria, which prompted the parents to seek medical attention.

It is not known why some eyes with vision loss become exotropic while others are esotropic or remain straight. Sidikaro and von Noorden found that in a series of 121 patients sensory esotropia and exotropia were found with almost equal frequency when the vision impairment occurred between birth and 5 years of age. Thereafter, exotropia was more commonly seen. von Noorden postulates that an individual's relative degree of tonic convergence may contribute to the direction of the sensory strabismus.[1]

Work-up

The first step in the work-up of a patient with esotropia must always be a thorough ophthalmologic evaluation, with an aim of detecting underlying ocular pathology which might contribute to vision loss.

Functional amblyopia must be considered in young children, as it is often superimposed on ocular pathology. Neutral density filters have proven useful in differentiating functional amblyopia from organic vision loss, as functionally amblyopic eyes will see better through a neutral density filter.[2,3]

Treatment

Treatment of esotropia secondary to vision loss related to ocular pathology is geared firstly to treating the underlying disease and restoring vision, wherever possible. Functional amblyopia is treated in young children. Treatment of the sensory esotropia is most often aimed at improving cosmesis through surgical correction. There is recent evidence to suggest that surgical re-alignment in such cases may also functionally expand an individual's visual field, even in cases of poor vision and absent fusion.[4]

ESOTROPIA SECONDARY TO DECOMPENSATED ESOPHORIA, MONOFIXATION SYNDROME AND SPONTANEOUS ESOTROPIA (BURIAN & FRANSCHETTI TYPE)

According to von Noorden the most common presentation of an acute onset, comitant esotropia occurs in individuals who experience temporary disruption of fusion while undergoing occlusion of one eye.[5] This may be associated with a medical reason for patching such as ocular trauma; following acute swelling of the lid(s); or after occlusion for anisometropic amblyopia. In the case of one of our young patients, a brief stint of uniocular occlusion while dressed as a pirate for Halloween was enough to precipate the onset of a constant esotropia.

Benign acute spontaneously decompensated comitant esotropia may also occur in children in the absence of occlusion. However, recent literature suggests that we must always carefully consider the presence of intracranial disease in such instances.[6,7]

Acute onset comitant esotropia may be secondary to an underlying esophoria and/or uncorrected refractive error. Often no cause for the decompensation can be pinpointed. Patients will usually demonstrate good binocular potential, and will respond well to treatment with resumption of fusion and stereopsis in the post-treatment phase.

Work-up

History will identify whether occlusion of one eye played a role in the decompensation of the esotropia. A cycloplegic refraction will detect hyperopia or anisometropia. Special care must be taken to rule out an esotropia secondary to an abducens paresis.

Treatment

Treatment of benign acute onset comitant esotropia in children is aimed at restoring comfortable BSV while maintaining good visual acuity either eye. Any hyperopia/anisometropia must be fully corrected, and amblyopia treated. Residual deviations generally respond well to strabismus surgery.

NEUROGENIC ACQUIRED ESOTROPIA

ABDUCENS PARALYSIS

Isolated abducens paresis or paralysis, with no other signs or symptoms, occur rather frequently in patients of all ages, particularly children. While these may be associated with vascular, inflammatory or compressive lesions, sixth nerve pareses are also seen in patients with systemic vascular disease, particularly hypertension, diabetes mellitus, cardiac disease or following vaccination or viral illness.[8] In a series of acute sixth nerve palsies in children, age 7 and under, Aroichane and Repka found the etiology and recovery rate to be as outlined in Table 2.[9]

While the etiology of a sixth nerve paresis/palsy in children may indeed be grave, Bixenman and von Noorden described a case of six episodes of recurrent acquired benign sixth nerve pareses in a child be-

TABLE 2[9]
ETIOLOGY OF AND RECOVERY FOLLOWING SIXTH NERVE PALSY OR PARESIS IN CHILDREN

Etiology	Number (%)	Recovery Rate (%)
Tumor	21 (33)	5
Hydrocephalus/ shunt malformation	15 (23)	33
Trauma	12 (19)	33
Infectious	4 (6)	50
Malformations	4 (6)	25
Idiopathic	3 (5)	67
Miscellaneous	5 (8)	20

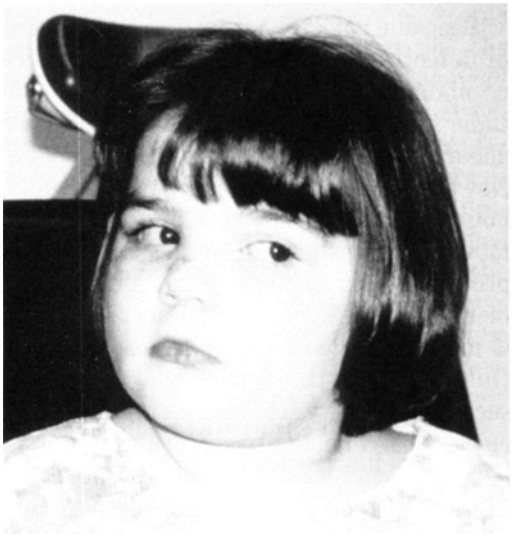

FIGURE 1: 5 y.o female with acute recurrent right VIth N paresis adopting a marked compensatory head posture-face turn to the right to achieve BSV at distance.

tween the ages of 2 1/2 months and 3 years.[10] Several others have reported similar cases.[11]

CASE 1:

Presumed benign recurrent VI N Paresis

A six year old girl was seen with a four day history of sudden onset horizontal diplopia and esotropia right eye. Her mother reported that she had experienced a similar episode one year previously, which had resolved completely and spontaneously within one week, while she awaited an appointment with an ophthalmologist. Her ophthalmic exam was completely within normal limits immediately following the first episode. A CT scan performed at that time was also within normal limits. The child was discharged, and the mother instructed to call immediately if the problem recurred. When seen in March, 1995, at the time of the second episode, the child was adopting a sizeable compensatory head posture, face turn to the right, to achieve binocular vision and avoid diplopia (Figure 1). She was otherwise healthy and on no medications. Specifically, there was no history of a recent viral illness or immunization. She did admit to a mild frontal headache. Visual acuity was 6/7.5+2 in either eye. Cycloplegic refraction was +0.50 OU. Pupils and fundi were within normal limits. A large, incomitant esotropia was noted at near and distance. The right eye was limited in abduction,

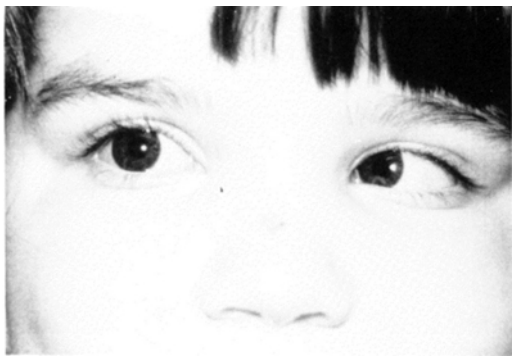

FIGURE 2: Same child as in Figure 1. Limitation of right eye on adduction.

but otherwise extraocular movements were full (Figure 2). Prism cover test measurements were as follows:

1/3 m pp	FR ET 25*
	FL E(T) 14
6m pp	FR ET 18*
	FL ET 12
6m gaze R	ET 45
6m gaze L	orthophoria
6m upgaze	ET 12
6m downgaze	ET 25

*It is interesting to note that in this case the esotropia fixing right eye, was larger at near than distance. In a sixth nerve paresis, one normally expects to see a larger deviation at distance than near.

Adopting a marked compensatory head posture, she achieved 140 secs of arc on Titmus, 550 secs of arc on Lang I and 200 secs of arc on Lang II stereotests. A repeat CT scan and MRI were both within normal limits. No active treatment was initiated. When the child was seen again twelve weeks later, the esotropia had resolved completely, and the extraocular movements were full. (Figure 3) According to her mother the esotropia resolved much slower the second time around, taking approximately eight weeks to return to normal. This is in keeping with other case reports which suggest that recovery is slower with each episode of benign recurrent VI N paresis in children.[11]

Work-up

Miller suggest that the most important factor in differentiating between a "be-

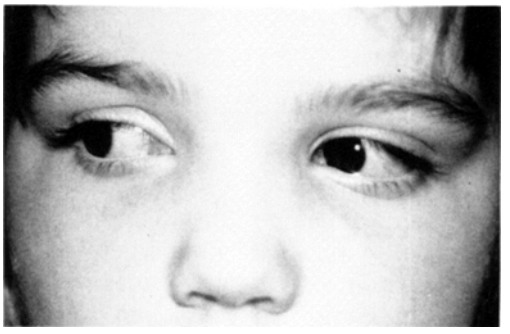

FIGURE 3: Same child as Figures 1, 2. Twelve weeks later right VIth N paresis resolved.

nign" sixth nerve paresis and one related to significant neurologic disease is the absence of other neurologic signs.[8] Miller suggests that all patients presenting with an abducens paresis undergo a thorough medical and neurological exam, with particular attention paid to other cranial nerves. If the sixth nerve paresis worsens or other neurologic signs develop, a thorough neuro-radiologic workup is suggested. Glaser advocates that children presenting with an isolated sixth nerve paresis and no other signs or symptoms, have a middle-ear infection ruled out, undergo at least plain skull x-rays including sinus views, and have a peripheral blood count carried out so as to obtain an indication of a recent viral infection.[12]

The diagnosis of an acute onset sixth nerve paresis is relatively easily made in most cases by the presenting history of uncrossed diplopia, worse at distance and to the affected side(s). Side gaze measurements should always be performed at distance fixation. To ensure maximum abduction of eyes for prism and cover measurement, one should turn the whole patient, not just his/her head, covering the abducted eye, allowing the patient to "just see" an interesting distant target with the adducted eye. Limitation on abduction may be very subtle or easily discernable, depending on the degree of paresis.

Saccadic velocity measurement is useful in detecting mild abducens paresis where no abduction limitation can be seen. Saccadic velocities are particularly useful for documenting recovery of paretic muscles. Newer technology utilizing goggles which house microchips for tracking limbal eye movements with infrared reflection shows promise as an important tool in the strabismus clinic.[13] (Figure 4) This technology is much faster and easier to use in young children than the traditional method of saccadic velocity testing using adhesive electrodes. The PC based system also allows for easier storage of recordings. However, our experience to date suggests that the software may require refinement to efficiently meet the needs of a busy pediatric ophthalmology and strabismus clinic.

Treatment

If a sixth nerve paresis is very mild, and the patient is satisfied to simply adopt a slight compensatory head posture to avoid diplopia and maintain binocular single vision, no active treatment may be necessary. Frequent follow-up is necessary to ensure that the paresis is not worsening, indicating a pathologic process. In children under

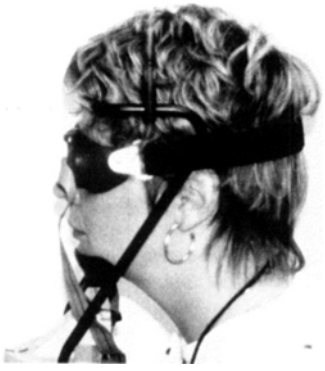

FIGURE 4: Ober2® Eye Movement Recording System utilizes goggles with microchips which track limbal eye movements by infrared reflection.

age 9, regardless of the degree of esotropia associated with an abducens paresis, the potential for amblyopia must be considered and carefully monitored. In most patients, symptomatic relief of diplopia resulting from an acute sixth nerve paresis can be achieved with Fresnel prisms, provided the eso-deviation does not exceed 40Δ, the maximum power available. As the esodeviation is usually greater at distance, often two base out prisms will have to be used, one for the upper portion of the spectacle lens and another as a "bifocal prism" for use at near. Due to the blur induced by the fresnel prisms, most patients prefer to have the prism put on one lens only. It is generally our practice to put the prism before the paretic eye, as this encourages fixation with the non-paretic eye, avoiding a secondary deviation. However, if the patient has a strong visual preference for the paretic eye, the prism may be placed on the other eye to achieve comfort. As the paresis recovers, the strength of the Fresnel prism can be gradually titrated, and eventually discontinued.

If the eso-deviation resulting from the abducens paresis exceeds 40Δ, patients will usually prefer to have the paretic eye occluded rather than wear fresnel prisms on both eyes. This can be achieved most comfortably with the use of a Bangerter foil adhered to a spectacle lens.

Many ophthalmologists use Botulinum A Toxin in the treatment of sixth nerve paralysis, injecting this into the medial rectus to prevent contracture as the abducens nerve function recovers. The dose of Botulinum Toxin cannot, however, be adjusted such that the eyes are aligned and BSV re-established immediately following injection. Rather, pre-treatment esotropia may become exotropia immediately following injection, with diplopia switching from uncrossed to crossed. Interestingly, in a randomized prospective trial designed to determine the prophylactic effect of Botulinum Toxin A in the treatment of acute onset abducens paresis, Lee et al found there to be no significant difference in the long term outcome between a Botulinum Toxin treated group and non-treated group.[14]

ACUTE ONSET ESOTROPIA SECONDARY TO INTRACRANIAL DISEASE

CASE 2:
Acute onset esotropia and diplopia in 7 y.o. female

A 7 year old female presented to our clinic with a 4 month history of acute onset esotropia and diplopia. Her sister had previously undergone strabismus surgery for esotropia. She was healthy except for seasonal asthma, for which she used a Ventolin® puffer as necessary. Her visual acuity 6/7.5 + 2 OD and 6/6-1 OS. Extraocular movements were full, in particular abduction was full OU. She reported uncrossed diplopia on W4D at near and distance. Pupils were normal, as were fundi and discs. History for intracranial disease was negative. She did not complain of headaches, nausea, vomiting, irritability or clumsiness. There were no other cranial nerve changes. Her prism cover test measurements were as follows:

1/3m pp	ET 40 with +3.00's ET 30-35
6m pp	FR ET 25 L HypoT 4 FL ET 30 RHT 4
6m upgaze	ET 20 RHT 6
6m downgaze	ET 30 RHT 5
6m gaze R	ET 30 RHT2
6m gaze L	ET 25 L HypoT 10
6m Head tilt R	ET 30
6m Head tilt L	ET 25 RHT 5

The vertical deviation suggested a left superior rectus paresis to the Three Step Head Tilt Test. There was no limitation of the LSR on extraocular movements, though she did require correction of the vertical deviation as well as the horizontal, to fuse on synoptophore testing. Fusion potential with a four degree convergence and two degree divergence amplitude range, but no stereopsis, was demonstrated at ET 40Δ RHT 6Δ on synoptophore testing. Her cycloplegic refraction of +1.00 OD and +0.75 was ordered and worn, but did not have a positive effect on the esotropia. Six

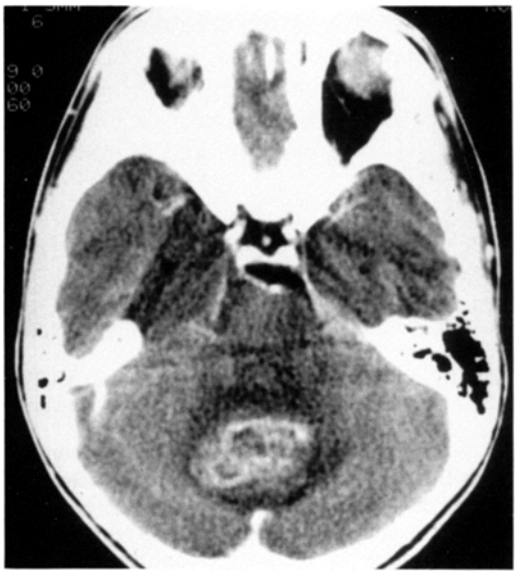

FIGURE 5: CT scan showing large posterior fossa tumor in 7 y.o. female who presented with sudden onset comitant ET and diplopia. No other neurologic signs present.

months later, there being no change, except that the diplopia was lessening somewhat, the child was scheduled for strabismus surgery. A CT scan was arranged for the day of admission. The surgery was cancelled when the CT scan revealed the presence of a large midline tumor of the posterior fossa. (Figure 5) The tumor was completely resected and diagnosed as a cerebellar astrocytoma. Within three months of the neuro-surgery, the esotropia was completely resolved and the child demonstrated 6/6 visual acuity in either eye, 40 secs of arc stereopsis on Titmus, 550 secs of arc stereopsis on Lang I and 200 secs of arc on Lang II. Two years later, she remains asymptomatic with straight eyes.

In 1989, Williams and Hoyt[7] described six patients ranging in age from three to ten years who presented with acute onset comitant esotropia, and who were subsequently found to have tumors of the brain stem or cerebellum. In each case the esotropia was the initial sign of the intracranial disease, and in no case did the esotropia become incomitant or manifest other signs of sixth nerve paresis. No papilledema or other sign of intracranial disease was initially apparent. The esotropia in these cases ranged from 15–35Δ. All patients complained of diplopia except one, a three year old who may not have understood what diplopia was. In one case, a +2.25 refractive correction was ordered, and this reduced the esotropia somewhat. None of the patients underwent neuro-radiologic investigation until further signs or symptoms developed, from three to seventy-one months following initial presentation. New signs included nystagmus in three cases, a mild bilateral facial paresis in one case and parental reports of poor motor coordination in two cases. Cerebellar astrocytoma was diagnosed in four cases, with one case of cerebellar medulloblastoma and one case of pontine glioma. Among the group of patients described by Williams and Hoyt none re-established fusion following resection of their tumors, and subsequent strabismus surgery. They recommended neuro-radiologic investigation in any patient in whom fusion is not regained following strabismus surgery for acute onset estropia.

Williams and Hoyt's extensive literature review describes case reports of acute onset esotropia also associated with hydrocephalus, shunt failure, Arnold-Chiari malformation and other instances of brain tumors.

More recently Astle and Miller[6] reported the case of a 15 year old male presenting with a six month history of acute onset comitant esotropia and diplopia. The esotropia measured 45Δ, was comitant fixing either eye in all positions of gaze. He also complained of headaches. There were no other neurological signs or symptoms, specifically no papilledema, nystagmus, pupil abnormality, nausea, vomiting or ataxia. A CT scan revealed the presence of a large posterior fossa tumor, which was subsequently determined to be a cerebellar astrocytoma. The major difference be-

tween this report and those of Williams and Hoyt was that this patient, like ours, regained fusion following tumor resection. Five years post-operatively he demonstrates an esophoria of 10–15 prism diopters and 40 secs of arc stereopsis on the Titmus test.

Workup

What can the clinician gleen from these case reports that would help identify acute onset estropia perhaps secondary to intracranial disease compared to benign decompensated esophoria or monofixation syndrome? The presence of associated headache, nystagmus, vertical deviation or other cranial nerve involvement are indicators of possible pathology and undoubtedly warrant further investigation. Astle and Miller strongly make the point that complaints of diplopia associated with acute onset comitant esotropia should always trigger further neurologic investigation.

DIVERGENCE PARESIS/PARALYSIS

Divergence paresis is a clinical entity consisting of acquired comitant esotropia and diplopia for distance which decreases or remains the same on side gazes, differentiating the condition from a sixth nerve paresis where the eso-deviation increases on side gaze. In divergence paresis, there is little or no eso-deviation at near, and ductions and versions are full. Fusional divergence amplitudes are markedly reduced or absent at near and distance. The literature points out that divergence paresis is almost always associated with intracranial disease, most often secondary to raised intracranial pressure arising from a variety of causes. It has also been associated with multiple sclerosis, encephalitis and lymphoblastic leukemia.[8]

Work-up

The diagnosis of a divergence paresis is made on the basis that fusional divergence amplitudes are absent at near and distance, even when fusion is reestablished with base-out prisms or at the patient's subjective angle on synoptophore testing. In addition, the esotropia at distance fixation is unchanged or decreases on side-gazes. Lim et al.[15] reported a slight reduction in abducting saccadic velocities (9–20 percent in 20 and 30 degree saccades) in a series of 12 patients with divergence paresis and no associated neurologic disease, as compared to normal subjects.

Treatment

Treatment of divergence paresis most often involves the use of base out prisms at least on a temporary basis. If the esotropia persists or prisms prove unsatisfactory, bilateral lateral rectus resection has proven effective.[16]

MYASTHENIA GRAVIS

Any discussion of acquired strabismus must include myasthenia gravis, the great imitator, in the differential diagnosis. Miller describes a 65 year old hypertensive man with no ptosis or other weakness who presented with an isolated abduction weakness of the right eye, which was presumed secondary to an abducens paresis. Within two weeks he developed ptosis and vertical diplopia and myasthenia was diagnosed.[17] Myasthenia gravis can mimic any isolated or multiple cranial nerve paresis. The diagnosis is usually made when the condition changes suddenly, significant variability in the angle of strabismus is noted, or if the patient presents with other symptoms of weakness.

Workup

Saccadic eye movements recorded before and after the injection of edrophonium chloride (Tensilon®) have proven useful in identifying myasthenia gravis. In our clinic, a Lees Screen is usually the diagnostic test of choice, and is plotted before and after Tensilon® injection. It provides a quick, consise and easily read endpoint and permanent record of any strabismus changes with Tensilon. Photographic records are also very useful.

ACQUIRED ESOTROPIA OF THE MECHANICAL TYPE

GRAVES (ENDOCRINE) OPHTHALMOPATHY

Though extraocular muscle involvement in Graves ophthalmopathy usually presents as a limitation of upgaze in one or both eyes secondary to tethering of the inferior rectus, esotropia combined with limitation of abduction secondary to tethering of the medial rectus is sometimes seen at the onset of the disease. Certainly it is common for the medial rectus to become involved as the disease progresses. While lid retraction, lid lag and exophthalmos usually make the diagnosis easily discernable, one occasionally sees a Graves patient without these other signs presenting as a pseudo-abducens paresis, with an acquired esotropia, diplopia and abduction limitation.

Work-up

The Lees/Hess screen is often very useful in differentiating strabismus which is restrictive from neurogenic in nature. An esotropia secondary to medial rectus tethering as in Grave's disease will show an eso-deviation which increases as the eyes attempt to abduct, but without corresponding overaction of the ipsilateral medial rectus.

Saccadic velocities may also prove useful in the differential diagnosis, and are usually normal or near normal in Graves disease. Forced ductions are positive and intraocular pressure will rise significantly on attempted abduction in Graves patients with tethering of the medial recti. Orbital ultrasonography and/or a CT scan will readily identify enlarged medial recti in these patients.

Treatment

Orthoptic treatment of patients with Graves ophthalmopathy usually involves the use of Fresnel prisms to eliminate diplopia. Graves patients are among the most satisfying for an orthoptist to treat, as they are so grateful for the symptomatic relief. While many Graves patients go on to strabismus surgery, that option is not usually considered until the deviation has proven stable for six months or more, and the endocrine imbalance is normalized with medical therapy. Patients with Graves disease must be watched carefully for visual acuity, contrast sensitivity and color vision changes which could signal optic nerve compression resulting from constriction by enlarged extraocular muscles.

MEDIAL WALL FRACTURE & IATROGENIC ACQUIRED ESOTROPIA

Iatrogenic esotropia secondary to medial wall orbital decompression carried out to preserve vision in a case of bilateral optic nerve compression in Graves ophthalmopathy is illustrated in Figures 6 and 7. Immediately following orbital decompression, the patient demonstrated a marked esotropia. This was surgically repaired with a good cosmetic and functional result. (Figure 8) A medial wall fracture secondary to any other trauma would result in a similar esotropia.

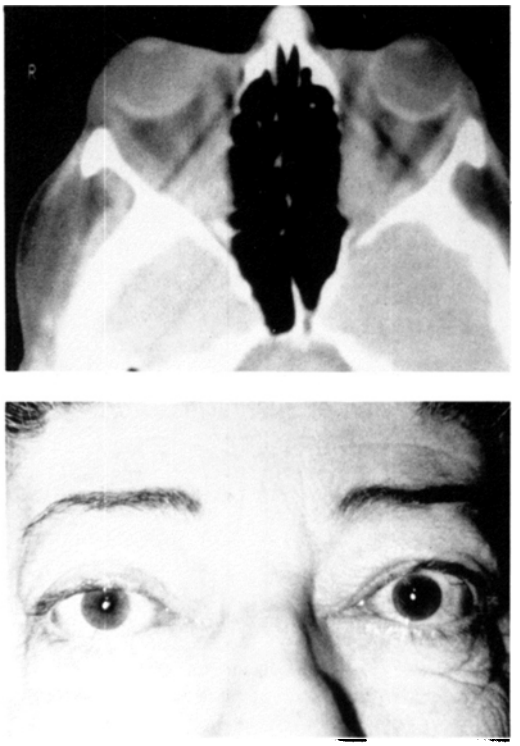

FIGURE 6: (Top) CT scan of patient with Graves Disease. There is marked enlargement of the medial recti OU resulting in optic nerve compression and decreased visual acuity. (Bottom) Same patient before orbital decompression.

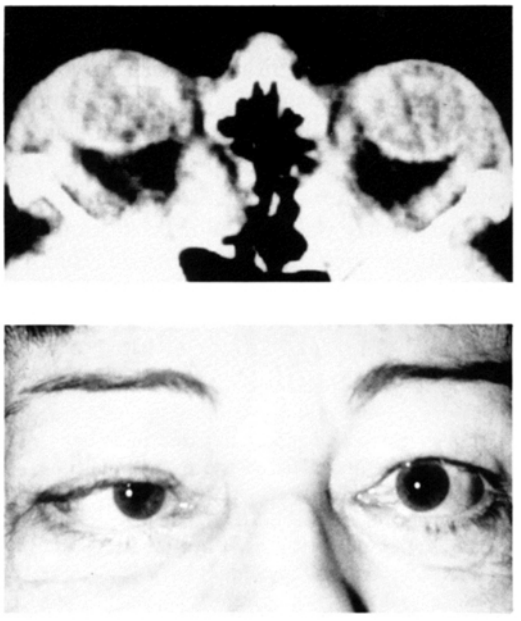

FIGURE 7: (Top) Same patient as Figure 6. CT scan after medial wall orbital decompression. The medial recti now occupy the ethmoid sinuses. (Bottom) The patient now demonstrates a marked (acquired) esotropia immediately following the orbital decompression.

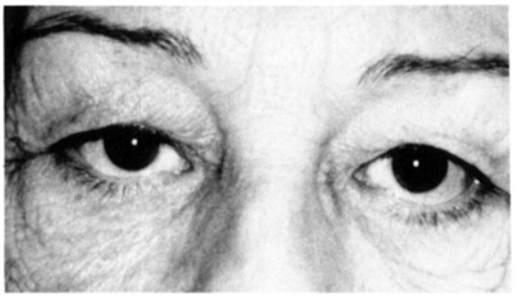

FIGURE 8: Same patient as Figures 6 and 7. After strabismus repair for ET; the eyes are straight.

ORBITAL MYOSITIS

Orbital myositis is a relatively rare inflammatory disease, with no known etiology, but presumably immunologic in nature.[18] Symptoms include an acute onset of pain, conjunctival injection over the insertion of the involved muscle, proptosis and diplopia, secondary to extraocular muscle inflammation.

Work-up

The diagnosis may not always be readily apparent, particularly in older individuals who, in our experience, do not necessarily experience pain as acutely as younger patients. A Lees screen will demonstrate a restrictive or mechanical problem rather than a paresis. A CT scan will illustrate acute inflammation of the involved muscle(s), and an orbital ultrasonography will confirm the diagnosis by the size of the extraocular muscle(s) as well as the relatively low reflectivity of the extraocular muscles compared to Grave's ophthalmopathy.

Treatment

Treatment of orbital myositis involves relatively high doses of systemic steroids, continued until the extraocular muscle(s) returns to normal size. Pain usually resolves soon after steroids are commenced, and frequently patients will be anxious to discontinue medication too soon. Recurrences are common.

CASE 4:
Orbital Myositis

A 72 y.o. physician presented with 14 day history of redness and swelling of his right eye, pain on eye movement and horizontal diplopia, worse at distance. He felt there was mild proptosis OD, and had also noted some nausea. His visual acuity was 6/7.5 OU. He had significant limitation of the right eye on abduction and mild limitation on adduction, and a slight esotropia in primary. (Figure 9). Exophthalmometry readings were OD 21.5mm; OS 19mm.

Prism cover test measurements were as follows:

1/3 m pp	FR, FL ET 12
6m pp	FR ET 16
	FL ET 12
upgaze	ET 12
downgaze	ET 12
R gaze	ET 25
L gaze	X(T) 10
Head tilt R	ET 20
Head tilt L	ET 20

Lees screen clearly showed a restrictive problem.

Ultrasonography and a CT scan confirmed inflammation of the right lateral rectus, (Figure 10) and the patient was immediately started on a course of systemic steroids. A 12^{Δ} base out Fresnel prism was adhered to the upper segment of his right spectacle lens and a 7^{Δ} prism fitted to the lower seg to relieve diplopia. Within two days of commencing the steroids the pain was gone. Steroids were continued until ultrasonography confirmed that the right lateral rectus had returned to its normal size, three months later. The diplopia resolved completely. Nine months following the initial presentation, the patient returned with complaints of similar ocular pain. Vertical diplopia was noted on extreme downgaze. Ultrasound and CT scan were normal, and the patient continues to be monitored carefully.

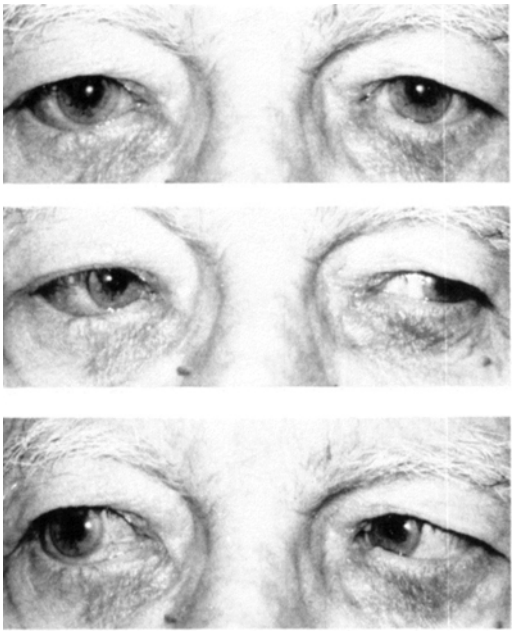

FIGURE 9: (Top) 72 y.o. male presenting with esotropia for distance fixation and orbital myositis involving the right lateral rectus muscle. (Middle) Conjunctival injection over insertion of the right lateral rectus muscle. He reported pain with eye movement. (Bottom) Limitation of abduction of the right eye.

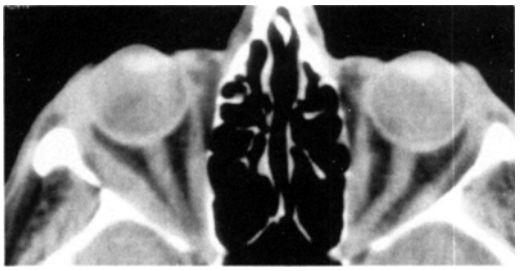

FIGURE 10: Same patient as Figure 11. CT scan reveals acute myositis involving the right lateral rectus muscle.

REFRACTIVE ESOTROPIA

According to Lang,[19] acute esotropia with diplopia, worse at distance, occurs occasionally in adults, particularly women, in whom myopic refractive errors are under-

corrected. This is apparently a rare phenomenon, seen in about one in one thousand cases, with a female to male ratio of four to one.

Von Noorden discusses another form of acquired esotropia, described mainly in the European literature, where the esotropia has a gradual onset and is associated with severe myopia, exceeding -15D. Both eyes may eventually become extremely esotropic, with positive forced ductions and limitations of extraocular movements.

We have occasionally seen patients, usually early presbyopes, present with clinical findings similar to a sixth nerve paresis, ie. sudden onset esotropia with diplopia—increasing at distance and on side gazes. Distance visual acuity is usually one or two lines less than one would expect given the remainder of the ocular exam. On cycloplegic refraction, significant *overcorrection* of myopia is detected. The mechanism of this entity is presumably an accommodative/convergence spasm. The esotropia resolves when the correct glasses are ordered, and have been worn for some time.

OTHER TYPES OF ACUTE ESOTROPIA

CYCLIC ESOTROPIA

Cyclic esotropia is a rare phenomenon characterized by straight eyes some days and large angle esotropia on other days. The rhythm follows a usual clock pattern, varying from 24 hours to 96 hours, depending on the individual. On straight days, patients usually demonstrate excellent binocular vision. Though more common in children, cyclic esotropia has been reported in adults.[8] Over time, it is reported that the clock mechanism eventually breaks down, and the esotropia becomes constant. Patients with cyclic esotropia generally do very well with strabismus surgery, aimed at correcting the maximum deviation. Patients usually regain full binocular function postoperatively.

VOLUNTARY ESOTROPIA

Voluntary esotropia is an interesting trick that some people can learn fairly easily. Occasionally, we have encountered children who have endured a complete neurologic and neuro-radiologic investigation because of the acute onset and diplopia associated with voluntary esotropia.

Workup

Pupils will be miotic in the "esotropic" phase. Individuals will be unable to sustain the voluntary esotropia when reading a small target at near with repeated cross cover testing. (Figure 11)

Treatment

Although children may be aware that they can voluntarily induce an esotropia, an over-reaction by a parent or health pro-

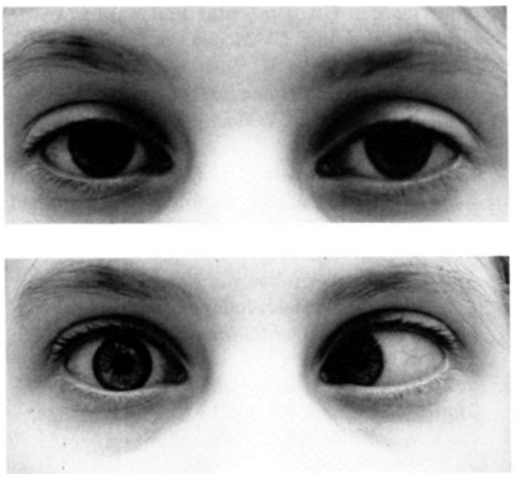

FIGURE 11: (Top) Eyes are straight, pupils are normal. (Bottom) Voluntary left esotropia. Note miosis of the pupils.

fessional can make them believe that indeed something is seriously wrong. The situation should be explained with tact and care. Usually the patient and her parents will be relieved that all is well.

CONCLUSION

Acute non-accommodative esotropia, in young children is most often benign. However, one should always rule out a posterior fossa tumor or other intracranial disease if any of the following are associated: nystagmus, vertical strabismus, other cranial nerve involvement, parental concerns of poor motor control, or persistent diplopia. Acute onset esotropia may also be the presenting sign of an ocular abnormality impairing visual acuity, a VI N paresis which may be associated with other disease entities, or a mechanical problem involving the extraocular muscles.

REFERENCES

1. von Noorden GK: *Binocular Vision and Ocular Motility.* St. Louis, CV Mosby Co. 1996, pp. 331–332.
2. von Noorden GK: *Binocular Vision and Ocular Motility.* St. Louis, CV Mosby Co. 1996, pp. 217–218.
3. Cadera W, Pachtman MA, Ellis FD, Helveston EM: Depth of strabismic amblyopia determined with neutral density filters. *Am J Ophthalmol* 1983; 95:763–766.
4. Wortham E, Greenwald MJ: Expanded binocular peripheral visual fields following surgery for esotropia, *J Pediatr Ophthalmol Strabismus,* 1989; 26:109–112.
5. von Noorden GK: *Binocular Vision and Ocular Motility.* St. Louis, CV Mosby Co. 1996, p. 324.
6. Astle WF, Miller SJ: Acute comitant esotropia: A sign of intracranial disease, *Can J Ophthalmol,* 1994, 29:3 pp. 151–154.
7. Williams AS, Hoyt CS: Acute comitant esotropia in children with brain tumors, *Arch Ophthalmol,* 1989, 107, pp. 376–378.
8. Miller NR: *Walsh and Hoyt's Clinical Neuro-ophthalmology* 4th Edition Volume 2, Baltimore, Williams & Wilkins, 1985, pp. 705–706.
9. Aroichane M, Repka MX: Outcome of sixth nerve palsy or paresis in young children, *J Pediatr Ophthalmol Strabismus,* 1995; 32:152–156.
10. Bixenman WW, von Noorden GK: Benign recurrent VI nerve palsy in childhood, *J Pediatr Ophthalmol Strabismus,* 1981; 18:329–34.
11. Knox DL, Clark DB, Schuster FF: Benign VI nerve palsies in children, *Pediatrics,* 1967; 40: 4560–564.
12. Glaser JS, *Neuro-ophthalmology,* 2nd ed, Philadelphia, J.B. Lippincott Co, 1990, p.369.
13. The Ober2 Eye Movement Recording System is marketed by permobil meditech Inc, 6B Gill Street, Woburn, MA 01801. The authors have no financial interest in the company.
14. Lee J, Harris S, Cohen J, Cooper K, MacEwen C, Jones S: Results of a prospective randomized trial of botulinum toxin therapy in acute unilateral sixth nerve palsy. *J Pediatr Ophthalmol Strabismus,* 1994; 31:5 pp 283–286.
15. Lim L, Rosenbaum AL, Demer JL: Saccadic velocity analysis in patients with divergence paralysis, *J Pediatr Ophthalmol Strabismus,* 1995; 32:2 pp. 76–81.
16. Krohel GB, Tobin DR, Harnett ME, Barrows NA: Divergence paralysis, *Am J Ophthalmol,* 1982; 94:506–510.
17. Miller NR: *Walsh and Hoyt's Clinical Neuro-ophthalmology* 4th Edition Volume 2, Baltimore, Williams & Wilkins, 1985, pp. 846–847.
18. von Noorden GK: *Binocular Vision and Ocular Motility.* St. Louis, CV Mosby Co. 1996, p. 450.
19. Lang J: *Strabismus,* Thorofare, N.J., Slack Publ. 1984, pp112–113.

Key words: acute onset esotropia, acquired esotropia, esotropia and pathology, myositis, intracranial disease and strabismus.