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A BRIEF REVIEW OF THE DIAGNOSIS AND TREATMENT OF INFANTILE ESOTROPIA

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Biography of Holly A. Parker

I am a Canadian from St. Albert, Alberta. I graduated from the University of Alberta in Edmonton, Alberta in June, 1987 with a Bachelor of Science degree. I became a member of the Beta Sigma Kappa Optometric Honors Society in 1988. In May, 1991 I received my Doctor of Optometry degree. I plan to return to Alberta to live and pursue my career objectives of joining a partnership or group practice and working in primary care optometry.

Abstract

The purpose of this review is to provide a brief overview of the literature as it relates to the differential diagnosis, testing, and treatment of infantile esotropia. The primary emphasis is on how all of these topics are approached when the patient is between birth and 24 months of age. Treatment strategy options for the practitioner are described with optometric therapy highlighted.

Vision care for infants and small children has become of increased concern to optometrists over the last decade. The scope and interest of the profession has been matched by advances in equipment and testing techniques which allow optometrists to be more skilled in handling the primary care needs of the youngest of patients. Infantile esotropia is reported as 28-54%[1,2] of all esotropia which must establish the significance of its diagnosis and management in providing prompt and appropriate care to these patients and their parents. Early treatment allows the maximum binocularity and visual potential to be developed.

Definition

Within the literature there is continuing debate about the exact terminology and definition for esotropia which has its onset during the first year of life. It is generally accepted that infantile esotropia is a broad, descriptive term used to identify a heterogeneous group of esotropes with an onset within the first year of life[1-3]. The terms congenital esotropia and essential infantile esotropia are considered by many to be equivalent[1-6] and define a specific esotropia which is manifest between birth and 6 months of age. The characteristics of essential infantile esotropia, some or all of which may be present, are listed in Table 1[2]. Some authors categorize esotropia with an onset after 6 months of age as 'acquired esotropia'[1,3].

Table 1. Characteristics of Infantile Esotropia

Angle of deviation between 30-60 PD
Alternation is common
Equal distance and near measurements
Inferior oblique overaction
Dissociated vertical deviation
Latent nystagmus
Accommodative component may be present
Absence of underlying disease

Diagnosis and Etiology

Esotropia in infancy may result from an hereditary predisposition or from a wide range of other causes. Table 2[2] lists the differential diagnosis necessary for determining the cause of the esotropia. The differential is important as some of the possible causes require medical referrals while others have less favorable

prognoses for functional cures even with all the current treatments available.

Table 2. Differential Diagnosis of Infantile Esotropia

1. Primary of essential infantile esotropia
a. Nonaccommodative
b. Accommodative
c. Partially accommodative
2. Esotropia secondary to underlying disease
a. Active pathologic conditions
b. Pathologic conditions that have stabilized and do not represent a health threat to the eye or child
3. Esotropia secondary to congenital syndromes
4. Esotropia associated with neurologic or multiple handicapping conditions
5. Pseudo-esotropia

Christenson et al[1] list and discuss pseudostrabismus, accommodative esotropia, sixth nerve palsy, Duane's retraction syndrome, Mobius' syndrome, and congenital esotropia in the differential diagnosis section of their article which provides a good background to the clinician. In his section on essential infantile esotropia, von Noorden's[4] differential includes sixth nerve palsy, dampening of manifest congenital nystagmus by convergence or nystagmus blockage syndrome, esotropia associated with congenital syndromes, refractive accommodative esotropia, sensory esotropia, and Duane's retraction syndrome. These are all well discussed in various sections of his textbook. An extremely well-done differential and discussion is provided by Scheiman and Wick[2]. In the section on esotropias secondary to underlying disease, the active pathology discussed includes encephalitis, meningitis, elevated intracranial pressure (hydrocephalus), retinoblastoma, and sixth nerve palsy. The stabilized pathologies given include complications associated with prematurity (ie. heterotopia of the macula), trauma, and injury. The congenital syndromes listed are Duane's syndrome, Mobius' syndrome and nystagmus blockage syndrome. Down syndrome, cerebral palsy, and menigiomyelocele (spina bifida) are discussed under the heading of esotropia associated with neurologic or multiple handicapping conditions. Psuedostrabismus is also detailed in a separate section. This provides an excellent outline for every clinician to follow in determining the underlying cause of the patient's infantile esotropia.

A brief discussion of the terms mentioned in the previous sections follows:

Pseudostrabsimus[2,7]: apparent strabismus due to an overhanging epicanthus which narrows the visible width of the sclera medial to the iris. A narrow interpupillary distance and a negative angle Lamda may also contribute to the appearance of esotropia.

Sensory esotropia[8]: results from reduced visual acuity in one eye which presents a barrier to sensory fusion and can result in esotropia. The origins are numerous, limited only by the number of pathological conditions which can affect the visual acuity of one eye. The most common causes are anisometropia, injuries, corneal opacities, congenital or unilateral traumatic cataracts, macular lesions, and optic atrophy.

Accommodative esotropia[9]: may be due to an abnormal demand on accommodation such as uncorrected hypermetropia or may result from the act of accommodating in association with a high AC/A ratio.

Dissociated vertical deviation[4,10]: is characterized by the spontaneous turning of either eye upward when the patient is fatigued or daydreaming or when fusion is artificially interrupted by covering one eye. The condition has been diagnosed in 51-90% of patients with infantile esotropia. It is infrequently present before 2 years of age and may develop years after satisfactory surgical alignment of the esotropia.

Cross-fixation[5]: is a manifestation in the majority of essential infantile esotropes where they alternate fixation in the primary position and cross fixate on side gaze, using the right eye in left gaze and the left eye in right gaze. The cross-fixation pattern can be confusing and mis-leading to the practitioner. The practitioner must remember that the absence of abduction so frequently encountered with these infants is secondary to the cross-fixation habit. These infants have never abducted an eye so it is impossible to evoke an abduction on either version or duction studies. Occlusion and the doll's head test are the diagnostic maneuvers for diagnosing or ruling out cross-fixation.

Encephalitis and Meningitis[2]: are inflammatory conditions generally caused by enteroviruses. Sequelae can include central nervous system dysfunction with involvement of intellectual, visual, auditory, and motor function. Parents will give a history of severe illness resulting in hospitalization.

Hydrocephalus and elevated intracranial pressure[2]: is a condition in which enlargement of the ventricular system develops from an imbalance between production and absorption of cerebrospinal

fluid. Treatment consists of a shunt between the cerebral ventricle and the peritoneum or the right atrium. If untreated it can result in brain damage and retardation. Usually ruled out in case history.

Retinoblastoma[2]: is the most common malignant tumor of childhood. It is often initially diagnosed by leukocoria (white pupil) caused by massive replacement of the vitreous by tumor which leads to sensory esotropia.

Sixth nerve palsy[2]: results when the sixth nerve or nerve nucleus is damaged or affected congenitally or by disease. It results in a noncomitant esotropia with an abduction deficit in the affected eye. If it is unilateral, the infant will adopt a compensatory head posture, with the face turned toward the eye with the paretic muscle. It is commonly associated with raised intracranial pressure and tumors.

Nystagmus blockage syndrome[2]: is characterized by infantile esotropia associated with nystagmus in abduction. The nystagmus is generally absent with the fixating eye in adduction but as the fixating eye abducts the nystagmus increases and the esotropia decreases. One characteristic finding is that when the infant is occluded, a face turn will occur to maintain the fixating eye in adduction.

Duane's syndrome[2,11]: an hereditary congenital syndrome in which the affected eye shows limitation or absence of abduction, restriction of adduction, retraction of the globe on adduction, narrowing of the palpebral fissure on adduction and widening on abduction, and deficient convergence. It is transmitted as an autosomal dominant trait. It is important to note that esotropia is not an essential component of the syndrome. The key finding in differentiating essential infantile esotropia from Duane's syndrome is comitancy testing.

Mobius' syndrome[2,12]: agenesis or aplasia of the motor nuclei of the cranial nerves characterized by congenital bilateral facial palsy in various combinations, with unilateral or bilateral paralysis of the abductors of the eye, sometimes associated with involvement of the cranials nerves, particularly the oculomotor, trigeminal, and the hypoglossal, and anomalies of the extremities. The differential diagnosis is not difficult due to the characteristic abnormal facial appearance and the likelihood of sucking and feeding problems associated with this syndrome.

Down syndrome[2,11]: a chromosome disorder chacterized by a small anteroposteriorally flattened skull, short, flat-bridged nose, epicanthal fold, speckling of the iris (Brushfield's spots), short phlanges, widened spaces between the first and second digits of hands and feet, small oral cavity/protruding tongue, and moderate to severe mental retardation.

Cerebral palsy[2]: it is a group of signs and symptoms that may be associated with many different diseases. The term "cerebral palsy" refers to any disorder of movement and posture that results from a nonprogressive abnormality or injury of the brain. Any injury to the brain during fetal, perinatal, or early development can lead to cerebral palsy. The difficulty in cerebral palsy lies in the brain's ability to control the muscles and nerves rather than a problem with the muscles and nerves directly.

Menigiomyelocele (spina bifida)[2]: is considered to be a congential malformation or deveopmental defect of the nervous system that occurs in the latter part of the first month of pregnancy when the spinal cord (myelo) and vertebrae around it do not form properly. This can result in paralysis and lack of function of lower limbs and organs, anomalies of the heart and bowel, and hydrocephalus. The history and physical appearance establish the differential diagnosis.

Testing Sequence

One possible testing sequence for approaching the differential diagnosis of infantile esotropia is given in Table 3.

Table 3. Testing Sequence

1. Case history and external examination of the eyes
2. Cover test and Hirschberg corneal reflex test
3. Occlusion, doll's head test, and EOM fields
4. Cycloplegic refraction
5. Dilated fundus examination

The case history would establish an age of onset for the esotropia, the constancy, developmental milestones, and any medical or family conditions - previous or on-going - which would indicate any pathologic or congenital causes. The external examination of the eyes would indicate any suspicions for pseudostrabismus such as wide epicanthal folds, any mongoloid or anti-mongoloid tilt which could indicate underlying congenital conditions and may be observed in some patients with A and V pattern strabismus[13], or leukocoria which would infer retinal or lens disease. The Hirschberg corneal reflex test and cover test would allow one to measure the magnitude of the deviation plus determine comitancy.

Occlusion may be needed for a short period (up to an hour) or for a longer period (1 to 2 days) in order to allow partial or full abduction to be manifest[1,2,4]. The doll's head technique is a

response where the eyes turn in the opposite direction of the head turn[1,2,4,14]. These tests can be used to rule out a sixth nerve palsy and also help establish whether or not cross-fixation (right eye used for left field of gaze and left eye used for right field of gaze)[5] is present. EOM fields will provide information on the action of all the muscles which would be beneficial in further distinguishing between the infantile esotropias.

A refraction and a cycloplegic refraction need to be done to determine the refractive error of the patient. Any possible accommodative component can be accounted for plus any astigmatism or anisometropia that might lead to amblyopia. The current philosophy is to prescribe the full plus found under cycloplegic examination[1,2,4]. The cycloplegic refraction should be repeated several weeks after the full plus has been prescribed to rule out any latent hyperopia.

A dilated fundus examination also needs to be performed. This will rule out any underlying retinal, optic nerve, or lens disease/condition which could be affecting the visual system and manifesting as infantile esotropia.

When testing it is pertinant to remember that essential infantile esotropia, accommodative esotropia, and nystagmus blockage syndrome result in comitant esotropias while noncomitant esotropias occur with meningitis, encephalitis, hydrocephalus, retinoblastoma, sixth nerve palsy, Duane's syndrome, Mobius syndrome, cerebral palsy, menigiomyelocele, and Down syndrome.

Treatment

Once the differential diagnosis has been made, the question for the clinician becomes, "What do I do now?" The answer depends in part on the underlying cause of the infantile esotropia. Table 4[1] provides favorable and unfavorable prognostic indicators for a functional cure via vision therapy/orthoptics. The table shows that

Table 4. Prognostic indicators for development of sensory-motor fusion via vision therapy/orthoptics

Favorable prognostic indicators	Unfavorable prognostic indicators
-later onset (after six months)	-early onset (first six months)
-short duration of condition	-long duration of condition
-moderate angle	-large angle
-intermittent strabismus	-constant strabismus
-concomitant	-non-concomitant
-absence of associated conditions*	-presence of associated conditions*
-normal correspondence (NC)	-anomalous correspondence (AC)
-presence of normal sensory-motor fusion at the objective angle in instrument (major amblyoscope)	-no sensory-motor fusion

* associated conditions e.g., amblyopia, double vertical dissociations, latent nystagmus, and overacting inferior oblique

essential infantile esotropia has a less favorable prognosis - a point stressed by von Noorden[4]. He does advocate early surgical intervention with prior correction of any refractive error (full amount of plus prescribed) or amblyopia and, as summarized by Press[3], divides his post-surgical results into the following classifications:

1. Subnormal Binocular Vision (Optimal Treatment Result)
   - Orthotropia or asymptomatic heterophoria
   - Normal visual acuity in both eyes
   - Fusional amplitudes
   - Normal Retinal Correspondence
   - Foveal supression in one eye in binocular vision
   - Reduced or absent stereopsis
   - Stability of alignment
2. Microtropia (Desirable Treatment Result)
   - Inconspicuous or no shift on cover test
   - Mild amblyopia frequent
   - Fixation central or parafoveal
   - Fusional amplitudes
   - Anomalous Retinal Correspondence
   - Reduced or absent stereopsis
   - Some stability of alignment
   - No further treatment except amblyopia prevention
3. Small Angle Strabismus (Acceptable Result)
   - Less than 20 prism diopters
   - Cosmetically acceptable
   - 80% have anomalous retinal correspondence
   - Less stability of angle
   - No further treatment except amblyopia prevention

4. Large Angle Strabismus (Unacceptable Result)
   Greater than 20 prism diopters
   Cosmetically unacceptable
   Less chance for ARC (supression prevails)
   Unstable angle

This classification system allows better "placement" of the patient by the clinician for discussing potential benefits and consequences of the treatment options with the parents.

Scheiman and Wick[2] advocate an approach that establishes the most rapid accurate alignment of the eyes via optical correction of the ametropia, additional plus power, incorportation of prisms, occlusion and active home therapy for treatment of amblyopia, suppression, and sensory/motor function, with surgery considered as a final option. Postsurgical care is designated as an absolute requirement in order to maximize binocular abilities. They also discuss the questions the clinician must be prepared to answer in presenting surgery as an option to parents - prognosis, best age for surgery, what surgery is best, the risks and benefits, and post-surgical care to list a few.

Christenson et al[1] provide a similar approach but present it in the form of flowcharts for those younger than 24 months old and those 24 months old and older. These are reproduced as Figures 1 and 2[1]. The sequence follows from monocular to binocular skills in vision therapy and also includes reference to post-surgical care if surgery is necessary. The gingham pattern mentioned in the flowchart indicates any sort of repetitive texture and can be easily done by draping a gingham pattern table cloth over the sides of the infant's play pen. This would create a patterned environment throughout the infant's visual world and stimulate the binocular cortical cells.

Press[3] summarizes the treatment approaches of Greenwald and Bateman et al in his review:

Greenwald - children ages 1 to 4:

1) Change the position of the infants's crib in the room so that the child is not presented with the same visual surroundings each day.
2) If the tropia is unilateral, feed the child toward the side of the turned eye so that visual contact is maintained.
3) Place toys or bright objects in the infant's path when creeping or crawling. If the toys are placed off center, the infant must use peripheral vision to locate them.
4) During periods of relaxed wakefulness, place the child on his belly with his head facing you so that the habitually fixating eye is occluded by the mattress. Use the sound of your voice or bright colored objects to maintain fixation.

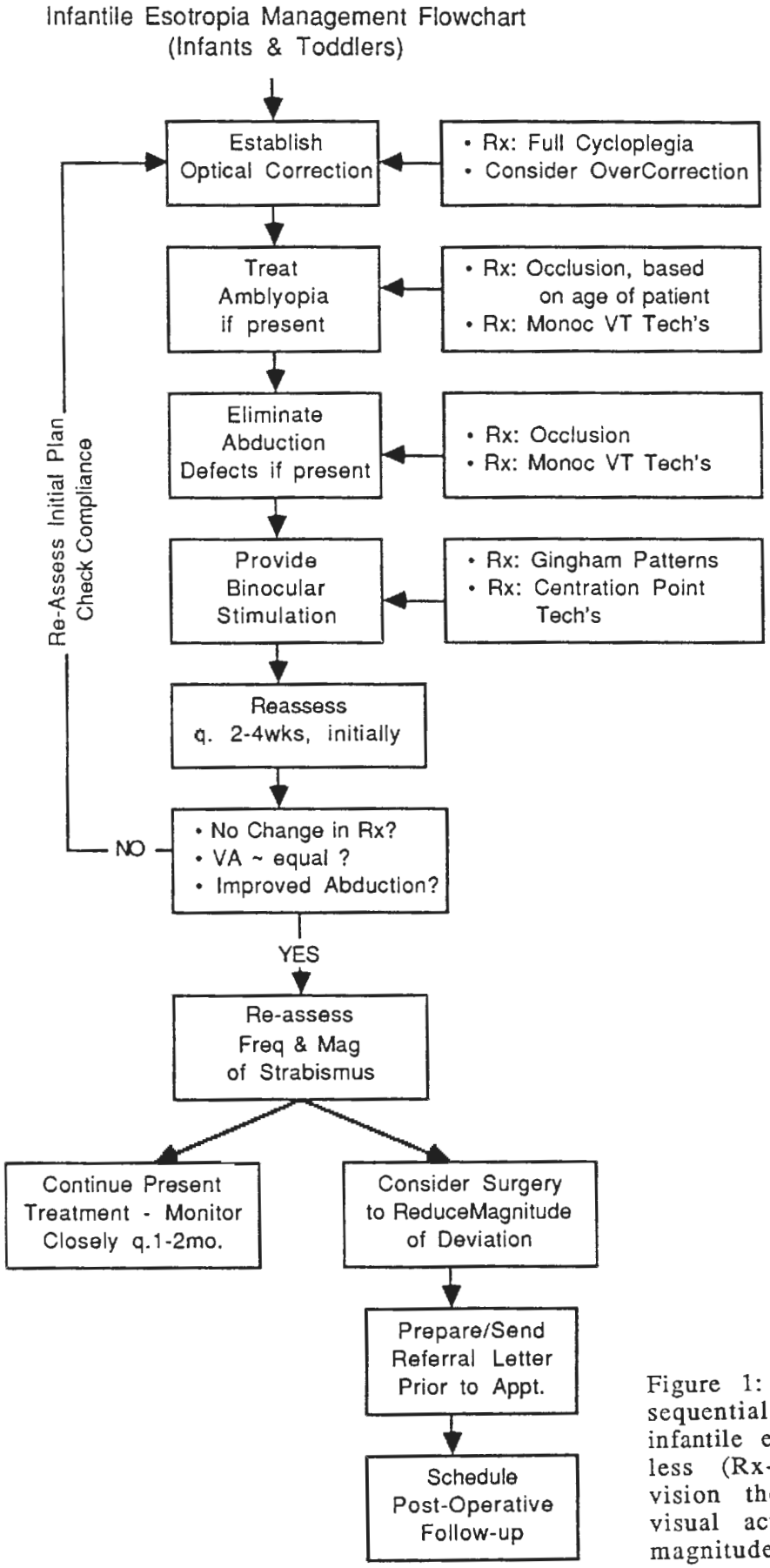

Figure 1: Flowchart emphasizing sequential management principles for infantile esotropes 24 months of age or less (Rx-prescription/prescribe, VT-vision therapy, q-each/every, VA-visual acuity, Freq-frequency, Mag-magnitude, Post-op-after surgery).

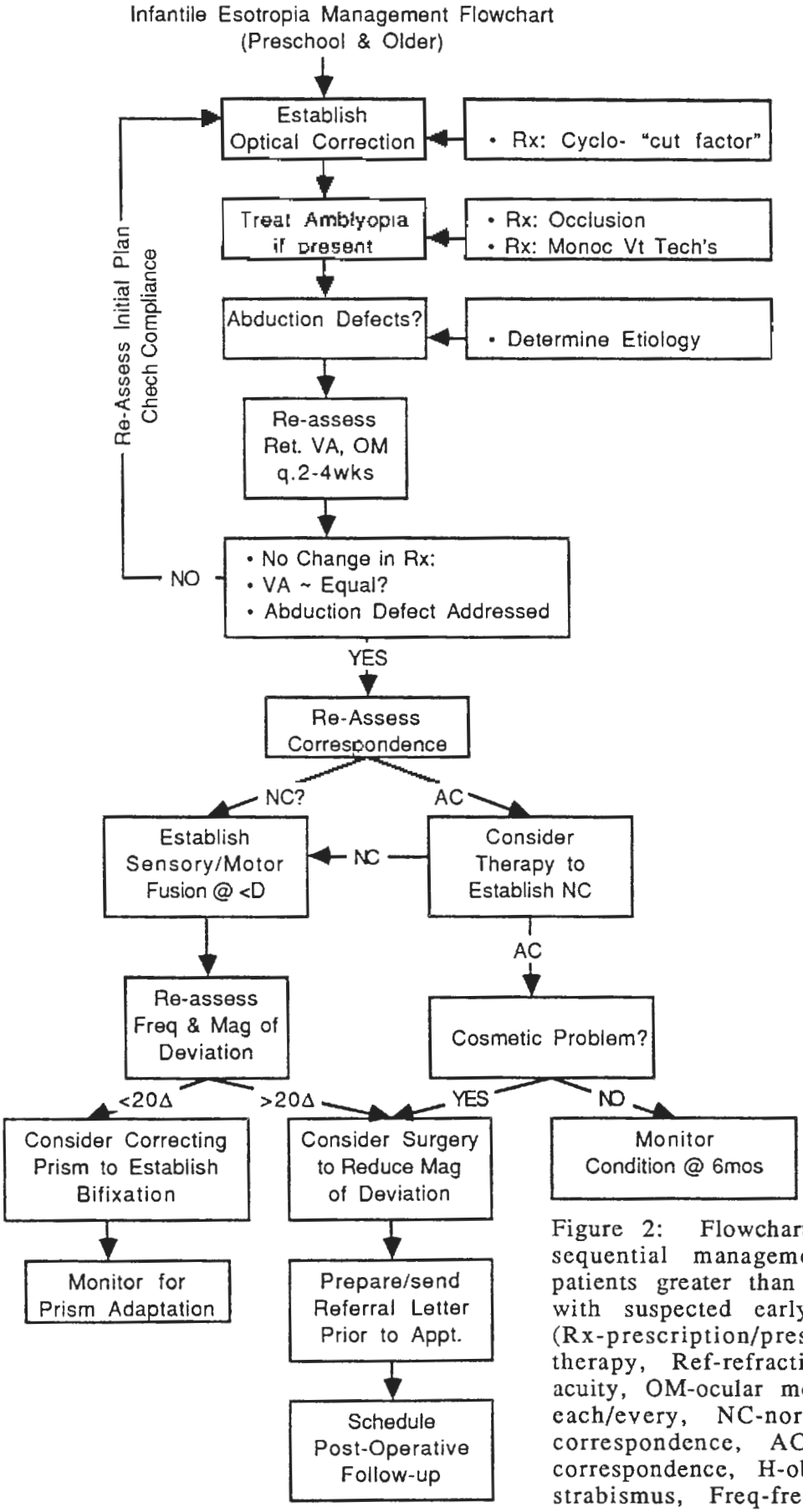

Figure 2: Flowchart emphasizing sequential management principles for patients greater than 24 months of age with suspected early-onset esotropia (Rx-prescription/prescribe, VT-vision therapy, Ref-refraction, VA-visual acuity, OM-ocular motility, q-each/every, NC-normal correspondence, AC-anomalous correspondence, H-objective angle of strabismus, Freq-frequency, Mag-magnitude, Post-op-after surgery).

5) Encourage hand-eye activity with items that encourage binocular visual integration such as pointer-in-straw or touch the dolly or placing small drinking cups inside larger cups, working one's way down from larger sizes to smaller sizes.
6) For unilateral esotropes, place narrow monocular tape on glasses on the side of the habitually non-deviating eye ("uni-nasal tape"). The width of the tape is gradually *increased*. The lenses are used for activities number 3 and 5 above.
7) Mobiles above the crib should be placed so that as they rotate, they are first seen by the turned eye.
8) Present gingham patterns for periods of 15-30 minutes per day. Theoretically this preserves overlapping of the two images and thereby impedes the development of ARC.
9) For constant esotropes, apply the exact amount of neutralizing prism to the glasses. 5-10 prism diopters of overcorrection can be tolerated. In unilateral strabismus the bulk of the Fresnell prism is placed before the habitually fixating eye. For 1-3 year olds, the prism is worn constantly until there is no further increase in the angle.
10) Monocular and/or binocular nasal occlusion can be coupled with the neutralizing prism in esotropia.

Bateman et al (summarized in three phases):
1) Equating Monocular Skills - includes gross motor, fine-motor postural conrol and bilateral integration, laterality and directionality, visual motor, and accommodative activities.
2) Fusion Development - includes physiological diplopia at the centration point, lustre, mirror superimposition, TBI, and visual/physical match with encroachment.
3) Fusion Range Extension - includes vergences ranges along the y and z axes, peripheral awareness, binocular rock, loose prism rock, directionality, spatial orientation, and ocular motor skills with base-in prism, and finishing techniques for binocularity such as BIM/BOP.

Both treatments sequences involve more wholistic approaches where the interaction with body/self and environment are stressed along with specific visual tasks to stimulate the strabismic eye(s).

Pharmacological agents can also be used in treating esotropia - particulary accommodative esotropia. The two parasympathomimetic (miotic) drugs that are routinely used are diisopropyl flurophosphate (Floropyl, DFP, 0.025% ointment) and echothiophate iodide (Phospholine iodide, 0.125% solution). Typically one drop of solution or one bead of ointment is instilled daily. von Noorden[15] has found that a 0.03% solution of Phospholine iodide is just as effective as a 0.125% solution in controlling the esotropia. von Noorden[4,15], Parks[5], and Griffen[16] all stress the fact that miotics must be considered as only a short-term measure in controlling esotropia and do not replace lens treatment. The side-effects and

complications include respiratory arrest if succinyl choline is used in general anesthesia, perspiration, nausea, vomiting, excessive salivation, frequent micturition, diarrhea, abdominal cramping, and development of iris cysts along the pupillary margin. Another drug that has been used is Botulinum A which is injected into the muscle while the patient is under general anesthesia. The treatment results in infantile esotropia have been found to be disappointing by von Noorden[4] and Biglan et al[17] which combines with Press'[3] list of complications - ptosis, secondary vertical deviations, persistant diplopia, perforation of the globe, and retrobulbar hemorrhage - to make one leery of considering this treatment option for infantile esotropia.

If the infant develops amblyopia, where one eye's visual acuity is less than the other, a period of occlusion therapy is warrented. The patching stimulates the amblyopic eye and the goal of therapy is to equalize the visual acuity and prevent the development of eccentric fixation. Eccentric fixation is an abnormality that is frequently observed in amblyopic eyes where the patient consistently directs a nonfoveolar retinal area toward the object of regard when viewing monocularly with the amblyopic eye[18]. The best evidence available indicates that conventional occlusion therapy is effective in eliminating eccentric fixation. The prevention of eccentric fixation is one reason for using a regime that involves direct and indirect patching. The general consensus in the literature is to use a patch that adheres to the skin for infants. von Noorden[19] suggests a 3:1 routine where the sound eye is patched for three days (direct patching), followed by the patching of the amblyopic eye for one day (indirect patching). Griffen[20] recommends a 2:1 pattern while Christenson et al[1] recommend a 1:1 routine. For these authors the term patching indicates that the patch is to be worn for the full waking day. Scheiman and Wick[2] advocate a multiple-periods-a-day patching sequence as long periods of occlusion can result in loss of visual acuity in the eye being occluded and a negative effect on binocular status. They recommend that the patch be used for 20-30 minutes three to four times a day with the patch placed on the child while the child is napping as the period right after waking up and being fed is the most desirable time for stimulating the amblyopic eye. Another occlusion technique is binasal occlusion[21] where the occlusive material (translucent tape, stippled clear nail polish, or black electrician's tape) is applied to the nasal portion of spectacle lenses extending temporally to some predetermined width. The material extends the full vertical length of the lens and may be tilted

inward 10 degrees inferiorally to allow for convergence. The temporal edge of the tape in front of the turned eye is placed so the corneal reflex is barely visible when both eyes are open while the temporal edge of the tape in front of the fixating eye is placed so it bisects the pupil. This encourages divergence of the turned eye while preventing or eliminating sensory maladaptions. The tape in front of the turned eye is moved temporally as the eye takes up a more aligned posture. Once the tape widths are the same they are gradually decreased in equal amounts as the patient is able to maintain the desired binocular status. All of the authors[1,2,19,20,21] stress that vision therapy techniques be used during occlusion to stimulate the amblyopic eye. These activities could include presentations of toys or other items with bright colors and sounds to attract monocular fixation, accommodation, and hand-eye coordination.

In total consideration, all of the treatment programs indicate the vast scope that vision therapy/orthoptics can play in the successful management of infantile esotropia and the importance of post-operative care to maximize the binocular potenial that can result after surgery.

Conclusion

The management of infantile esotropia is within the realm of optometric care. The skills and knowledge to deal with the differential diagnosis and treatment are developed during one's training. Press[3] lists these five steps as being the minimum necessary for proper management of infantile esotropia:

1) proper diagnosis and referral when appropriate
2) appropriate follow-up care
3) informing the parent of the condition
4) presenting the options available
5) explaining what to expect with and without treatment

In the years to come, optometrists will continue to expand their role in the vision care of infants and young children and should feel confident in approaching these patients with the research and recommendations of their colleagues to assist in the proper management.

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