UNUSUAL PRESENTATIONS OF ACCOMMODATIVE ESOTROPIA*

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ABSTRACT

Purpose: Most patients with accommodative esotropia are first examined between the ages of 6 months and 2 years. This paper discusses unusual presentations of accommodative esotropia that occur outside of this age-group and/or have a precipitating event that triggered the esotropia. In a series of patients who were from 5 to 11 years of age, trauma was the precipitating event. In some of the patients under 6 months of age, high myopia, as well as a moderate to large amount of hyperopia, was the cause. In 1 teenager, diabetic ketoacidosis precipitated accommodative esotropia.

Methods: We reviewed all of our records for the past 25 years involving patients with a diagnosis of esotropia, and we found 17 patients who had unusual presentations of accommodative esotropia. Of 8 who were under the age of 6 months, 2 had high myopia and 6 had moderate to large amounts of hyperopia. Nine patients were older than age 5. Eight of the 9 had suffered trauma associated with the presentation of accommodative esotropia, and 1 patient's accommodative esotropia was associated with diabetes. The patients with myopia received their full myopic correction. The children under 6 months of age with hyperopia received their full cycloplegic refraction, and the children over age 5 received the most plus that they were able to accept in a noncycloplegic state consistent with good visual acuity (at least 20/30 in each eye).

Results: In 17 patients, accommodative esotropia was initially controlled with glasses. In a few of the trauma cases, bifocals were required for control of near deviation. Only 2 of the patients, in whom onset was under 6 months of age, came to surgery. One had hyperopia controlled for 2 years with glasses, and the other had myopia controlled for 3 years with glasses.

Conclusions: Accommodative esotropia can occur prior to 6 months of age. It can also occur in older children (5 to 14 years of age) and can be precipitated by trauma or diabetic ketoacidosis.

Tr Am Ophth Soc 2000;98:119-125

INTRODUCTION

Accommodative esotropia is a common problem that ophthalmologists see in children. It occurs most often between the ages of 6 months and 2 years. However, it can occur in children up to 7-8 years of age.[1,2] Accommodative esotropia has also been reported in infants under 1 year of age[3] and even in a few infants under 6 months of age.[4] This study reports on a series of 17 children who had unusual presentations of accommodative esotropia. Some of our patients were only a few months old at presentation and were corrected with glasses. Others presented for the first time between the ages of 5 and 11, and their esotropia was precipitated by trauma. In a teenager whose diabetes was previously undiagnosed, onset of accommodative esotropia was concurrent with diabetic ketoacidosis.

The purpose of this paper is to alert ophthalmologists to these unusual presentations so that the appropriate diagnosis will be considered and the proper treatment by optical correction of the refractive error can be offered. We present the long-term follow-up results for these patients.

METHODS

We reviewed the records of all patients in our practice from 1974 to 1999 who had a diagnosis of esotropia. We found 17 patients who had unusual presentations of accommodative esotropia. Group 1 consisted of 8 patients under the age of 6 months. Included in this group were 6 patients with hyperopia varying from +3.50 diopters (D) to +6.00 D. The youngest was 3 months old when first examined and the oldest 4½ months. The amount of esotropia was the same for distance and near in all patients in this group and varied from 16 prism diopters

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(PD) to 50 PD. Two patients younger than 6 months of age presented with esotropia and 8 D of myopia in each eye. Initially, both were successfully treated with glasses; 1 eventually required surgery.

GROUP 1

Patient 1

A 3-month-old girl presented with 45 PD of esotropia at distance and near with a refractive error of +4.50 in each eye. The mother stated that the patient's eyes had been crossing for 2 weeks. Otherwise, the girl's neurologic development was normal. Findings on the remainder of the eye examintion were normal. During the examination, the patient had an intermittent esotropia varying from orthophoria to 45 PD of deviation at distance and at near (Fig 1). The full cycloplegic error of +4.50 was given to both eyes. At follow-up examination 1 month later, the patient had orthophoria at distance and near (Fig 2). After 13 years, her esotropia is fully corrected with glasses.

Patient 2

A 4-month-old girl presented with 16 PD of esotropia at distance and near with a refractive error of +4.00 in each eye. Her parents had noted an intermittent esotropia for 6 weeks, which had become constant. Her neurologic development had been normal. Glasses were given, and 1 month later the patient presented with orthophoria at distance and near while wearing her glasses. She remained controlled for 2 years, when she presented with esotropia of 25 PD at distance and near with her glasses. Without her glasses, she had 40 PD of esotropia at distance and near. A cycloplegic refraction showed her to still be hyperopic at +4.00 in each eye. She underwent a bimedial recession of 4.0 mm, and a 10-year follow-up showed her esotropia to be controlled with her glasses. While wearing her glasses, she had no deviation at distance and near, but without her glasses she had 20 PD of esotropia at distance and near.

Patient 3

A 4½-month-old boy with a history of eyes crossing for 2 weeks presented with esotropia of 45 PD at distance and near (Fig 3). The parent stated that initially the deviation had been intermittent, but at the time of the examination, it was constant. Refraction showed –8.00 in each eye, which was prescribed. At examination 1 month later, the patient had orthophoria at distance and near with his glasses (Fig 4). At a 10-year follow-up examination, his esotropia was well controlled at –10.00 in each eye (Fig 5).

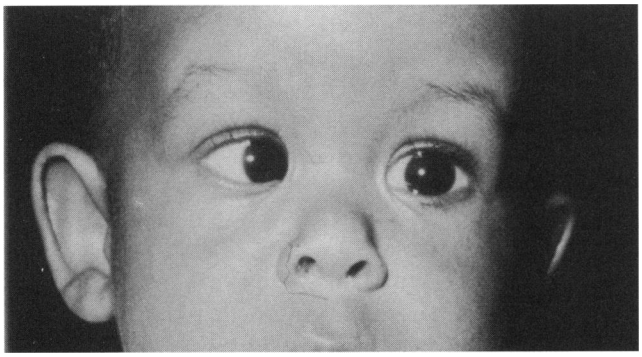

FIGURE 1
Patient 1. At 3 months of age, she presented with 45 PD of esotropia.

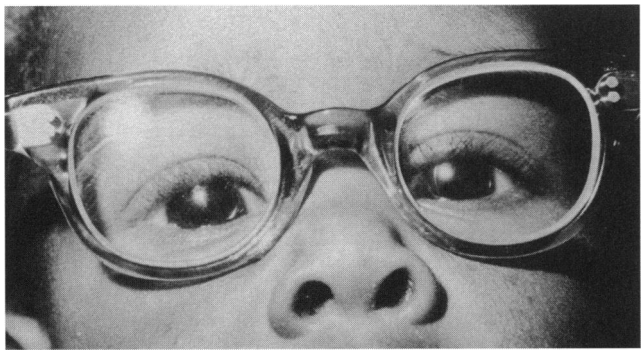

FIGURE 2
At 4 months of age, patient 1 had orthophoria with glasses.

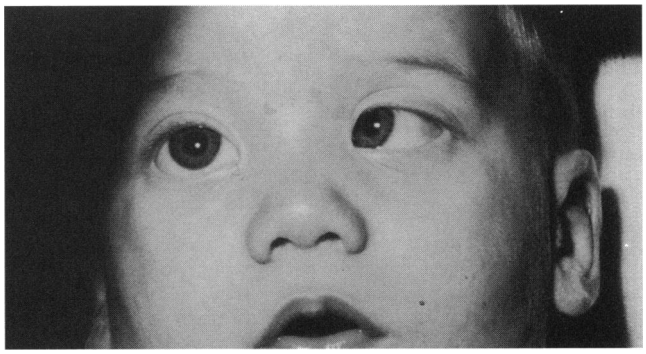

FIGURE 3
Patient 3. At 4½ months of age, he presented with 45 PD of esotropia.

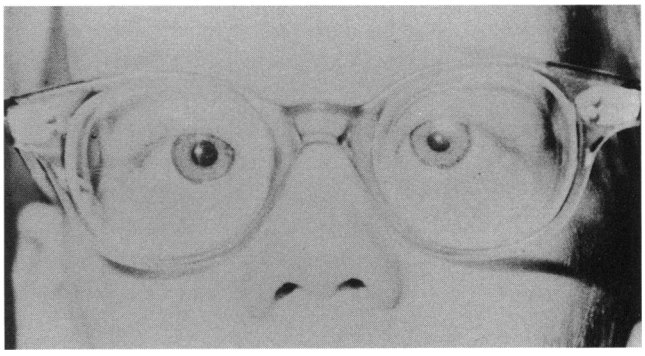

FIGURE 4
At 5½ months, patient 3 had orthophoria and –8.00 refraction in both eyes with glasses.

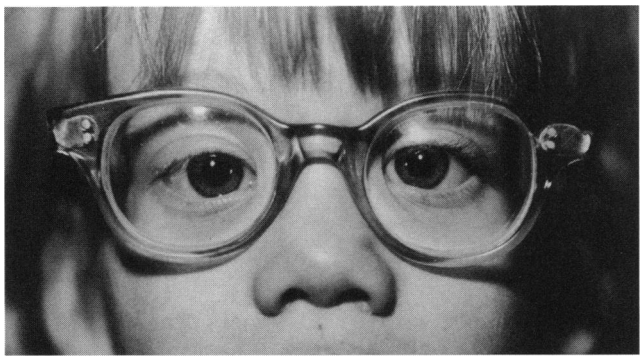

FIGURE 5
At 10 years of age, patient 3 had orthophoria with -10.00 refraction in both eyes.

GROUP 2

Group 2 consisted of 8 children whose accommodative esotropia developed between 5 and 11 years of age. Onset was secondary to trauma.

Patient 4
A 6-year-old boy who had been attacked by a dog had an injury to his left eye. He presented with 30 PD of esotropia in the primary position at distance, with orthophoria to the right, 60 PD of esotropia to the left, and 25 PD of esotropia in the primary position at near. There was no abduction of the left eye past the midline. The patient was taken to surgery, where the left lateral rectus was found to be 7 mm posterior to the insertion. The muscle was reattached to its original insertion. Two weeks later, abduction was full, but the patient had a comitant esotropia of 35 PD in all fields of gaze at distance and near. Cycloplegic refraction was +6.00 in each eye, but the patient would accept only +5.00 in each eye in a postcycloplegic state. One month later, he was orthophoric at distance and near with his glasses, and a 3-year follow-up examination has shown his condition to be stable.

Patient 5
A 5-year-old boy presented with a scissors injury to his left eye. His cornea, which had been lacerated, was repaired with 10-0 nylon sutures, which were removed at 3 weeks. He subsequently developed esotropia of 20 PD at distance in all fields of gaze and 40 PD at near. His refraction was +2.50 OD and +3.50 +3.00 x 050 OS. The full cycloplegic refraction with a +2.50 bifocal was given, and full-time patching of the right eye was started. Two months later, he presented with orthophoria at distance and 20 PD of esotropia at near, which reduced to orthophoria at near through the bifocal. At a 2-year follow-up examination, his vision was stable with his glasses, but he still had amblyopia in the left eye with a visual acuity of 20/60.

Patient 6
An 8-year-old girl presented with a sixth nerve palsy in the right eye after being knocked unconscious in a car accident. She had 50 PD of esotropia in the primary position at distance (Fig 6) with orthophoria to the left (Fig 7) and 60 PD to the right. She could abduct the right eye only 20 degrees past the midline (Fig 8). At near she had 30 PD of esotropia in the primary position. Over the next 3 months, her sixth nerve palsy resolved, but she was left with 45 PD of esotropia at distance and 65 PD at near. Her cycloplegic refraction was +6.00 in each eye, but she would accept only +4.00 in each eye in a noncycloplegic

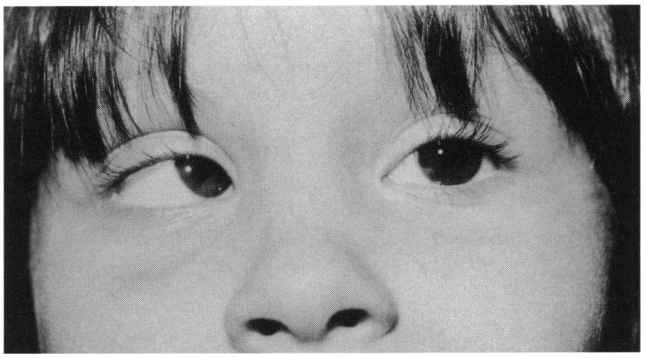

FIGURE 6
Patient 6. She had 50 PD of esotropia in the primary position at distance.

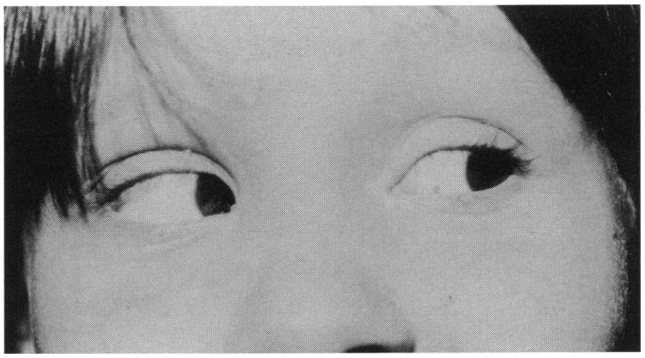

FIGURE 7
Patient 6. Orthophoria is present on gaze to the left at distance.

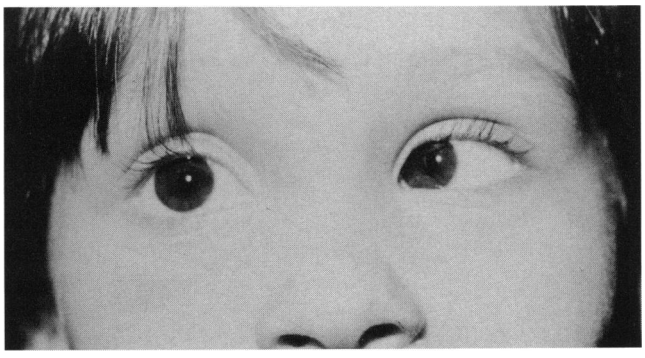

FIGURE 8
Patient 6. Right eye has limited abduction.

state. She was given glasses that were +4.00 in each eye with a +2.50 add. Two months later, she presented in her glasses with orthophoria at distance (Fig 9) and esotropia of 35 PD at near (Fig 10). The near deviation reduced to orthophoria at near through the bifocal (Fig 11). At a 5-year follow-up examination, her vision was well controlled with her glasses.

Patients 7 through 11

For the other 5 patients in this group, trauma was also the initiating event for onset of accommodative esotropia. An 11-year-old boy had a 60% anterior-chamber hyphema secondary to a hockey puck injury. The hyphema cleared in 7 days. He developed an accommodative esotropia within 2 months of the injury. The esotropia has been controlled with glasses for 5 years.

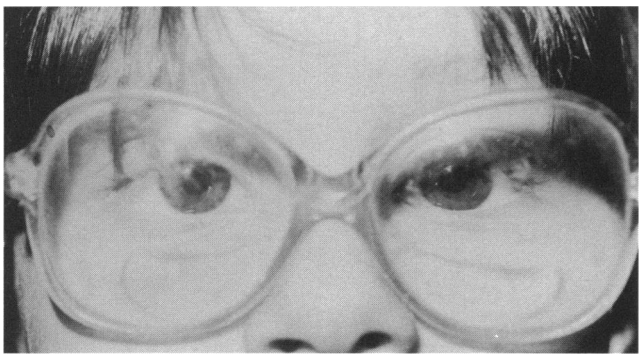

FIGURE 9
Patient 6. Orthophoria is present at distance with glasses.

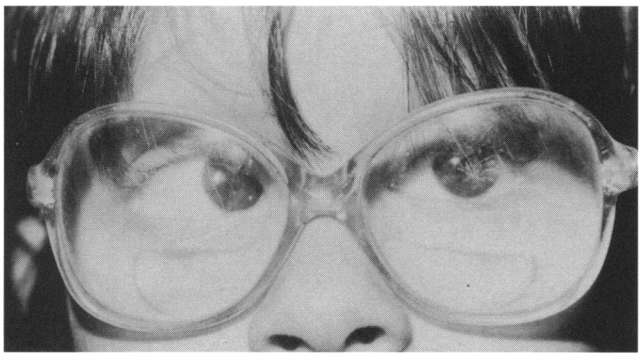

FIGURE 10
Patient 6. Esotropia of 35 PD is present at near

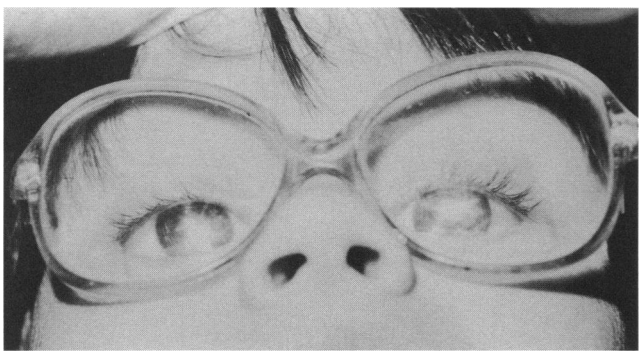

FIGURE 11
Patient 6. Orthophoria is present at near through bifocals.

A 6-year-old boy presented with an acute lateral rectus palsy. The lateral rectus muscle had been severed by a cat's claw. The severed muscle was repaired 4 hours after the injury with an end-to-end anastomosis. The palsy totally cleared 2 months after surgery. An accommodative esotropia developed 2½ months after surgical repair of the severed muscle, and this has been controlled with glasses for the past 7 years.

The other 3 patients in this group developed a sixth nerve palsy. One palsy was associated with a viral illness, and the other 2 were the result of head trauma received in car accidents. In all 3 cases, the palsy resolved, but the patients were left with an accommodative esotropia, which was treated with single-vision glasses. Follow-up for these 3 patients has been 5, 7, and 9 years, respectively, and their accommodative esotropia is under excellent control with glasses.

GROUP 3

This group consists of only 1 patient, who was 14 years old when she presented with a comitant esotropia of 60 PD in all fields of gaze at distance and near. She was quite ill with diabetic ketoacidosis but was alert and able to be examined. Her diabetes had been previously undiagnosed. Cycloplegic refraction showed +2.75 in each eye, but she would accept only +1.75 in each eye. She was given glasses, which she wore for 6 weeks. She was orthophoric at distance and near with her glasses. Six weeks later, she had relatively good control of her diabetes and no longer required her glasses for control of her esotropia. In 2½ years of follow-up, her esotropia has not recurred.

RESULTS

All 17 of these patients responded to glasses with control of their deviation. However, 2 patients eventually decompensated and required surgery. One was a 4-month-old boy with –8.00 in each eye whose esotropia was controlled by glasses until age 3, when he developed esotropia of 30 PD at distance and near with his glasses. He responded well to a unilateral medial rectus recession and lateral rectus resection. The other was a 4-month-old girl with accommodative esotropia controlled with +4.00 in each eye. At 2½ years of age, she developed a nonaccommodative component and underwent a bimedial recession, which controlled the nonaccommodative component of her esotropia. She has been controlled with use of contact

lenses and has been under observation for 10 years.

The children under age 6 months were given the full cycloplegic refraction. The 2 with myopia were given the full myopic refraction. The patients over 5 years of age were given the maximum "plus" that each would accept consistent with relatively good vision of at least 20/30 in each eye. This usually meant giving less than the full cycloplegic refraction. (This was always measured in a noncycloplegic state.)

DISCUSSION

In 1958, Parks[5] noted that there were 3 origins for accommodative esotropia: hypermetropia, a high ratio of accommodative convergence to accommodation (AC/A), and a combination of the 2. The average age at onset was 2½ years. In those patients with a normal AC/A ratio, an average amount of hypermetropia of +4.75 was seen. The patients with a high AC/A ratio had an average hypermetropia of +2.25. Our patients ranged from +3.50 to +6.75. Three of our patients with trauma as the precipitating factor did require bifocals.

Costenbader, in 1961,[6] stated that infantile esotropia is any esotropia noted before the age of 1 year. He changed his statement in 1968,[7] when he said that the more exact term for early-onset esotropia was congenital esotropia, which he defined as esotropia with onset prior to 6 months of age. It is obvious from this study that accommodative esotropia can have an early onset, even before 6 months of age. Children at this early age do appreciate the gain in vision afforded by their accommodative efforts. We usually do not include children with congenital esotropia in the same group as children with accommodative esotropia. A commonly held belief is that congenital esotropia is nonaccommodative. If we consider that congenital esotropia is esotropia occurring prior to 6 months of age, where do we put the patients with accommodative esotropia that occurs before age 6 months of age? They should be considered as a entity separate from congenital esotropia. Where do we classify the 2 patients with high myopia and accommodative esotropia? These young children cannot make their vision better by any means, so they overaccommodate to try to clear their vision. Obviously, this attempt makes their vision worse, but when their vision is cleared with glasses, they stop accommodating and their esotropia is eliminated.

Von Noorden[3] recommended that all hypermetropic errors in excess of +2.00 be corrected before surgery is considered. We also correct all errors in excess of +2.00 in children with acquired esotropia who are over 6 months of age. In patients under 6 months of age, we have not corrected any errors lower than +3.50 in the past 15 years. Prior to that time, we treated some children under 6 months of age with errors of +3.00. Glasses had no effect for these patients.

The amount of deviation should not deter one from making a diagnosis of accommodative esotropia. Usually, accommodative esotropia has a deviation from 20 to 40 PD,[8] but 1 of our patients had only 16 PD, and 1 had 50 PD.

The children with trauma had no prior history of strabismus. The fusional mechanism was upset with the trauma, which allowed the hyperopic demands to set the stage for accommodative esotropia. Even after the traumatic event had passed and the ocular motility had returned to normal in those with a lateral rectus palsy, their esotropia could not be controlled without the help of glasses. One patient had just suffered a hyphema with no lateral rectus muscle palsy.

The vision of the teenager who had diabetic ketoacidosis and accommodative esotropia was restored to normal when her metabolic state was under control. We have seen many patients with neurologic insults who develop esotropia. The majority regain control of their deviation within 3 to 4 months after the neurologic insult has passed. These are patients with shunt failures, pseudotumor, or concussions. The diabetic teenager described here is the only one we have seen with this presentation in 25 years of practice.

CONCLUSION

Children younger than 6 months of age do accommodate, and therefore accommodative esotropia can occur in this age-group. A diagnosis of accommodative esotropia should be considered in children with moderate amounts of hyperopia (at least +3.50) who present with esotropia in the first 6 months of life. Intermittence of the deviation may also be a clue to the diagnosis. The amount of deviation should not deter one from making this diagnosis, because we have seen deviations as small as 16 PD and as large as 50 PD.

The children described here who had trauma and the teenager who had diabetes should alert us to the possibility of accommodative esotropia in older children. If a significant amount of hyperopia is present (all of these had at least +2.50), then an accommodative etiology should be considered, even when there are other circumstances, such as hyphema, acute traumatic or viral sixth nerve palsy, or diabetic ketoacidosis.

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DISCUSSION

DR DAVID R. STAGER. I would like to congratulate Drs Pollard and Greenberg on a very interesting paper and thank them for supplying their manuscript in a timely manner for my review. I think it stresses to us the importance of always being cognizant of the possibility of accommodative esotropia, even in the non typical age group. I am amazed at their ability to review 25 years of medical records to identify these unusual cases. I am hopeful that those of us without computer minds will someday have access to this type of research once computerized medical records become available. I would like to comment on the 2 groups they have addressed.

First the infantile group. One common error they avoided is relying on parent history when ascertaining the onset of deviation in children. That can be notoriously inaccurate. Their definition is based on their observation of the problem under 6 months of age. These patients all have a moderate and intermittent esotropia which is a major characteristic of accommodative esotropia. We know from a recently published report by Eileen Birch and the congenital esotropia observation study group that infants with large angle esotropia at 3-6 months of age almost never straighten spontaneously, even with significant hyperopia corrected with glasses. Still, we usually prescribe the hyperopic correction when the refractive error is 3 diopters or more, even in small infants. This is particularly important since accurate measurement of esotropia is difficult at best in these infants and what may look to be 40 diopters may actually measure closer to 60 or 70 diopters on cross cover testing. I wish to ask whether these patients have the DVD, latent nystagmus, motion asymmetry and sensory outcomes that are characteristic of congenital esotropia or findings more characteristic of accommodative esotropia acquired after 1 year of life.

Regarding the older group (5 years or more), the lesson here is that anything which disrupts fusion, be it VI nerve palsy, patching, anisometropia or uncorrected hyperopia or overcorrected myopia, may precipitate esotropia. We see late onset accommodative esotropia also in patients in their 30's who are not appropriately corrected. Thus we agree that late onset esotropia needs to have adequate cycloplegic refraction with an appropriate refractive adjustment. An additional question is whether you reduce a full correction until the child has 20/30 vision or do you prescribe the least amount of plus that will enable the child to control the esodeviation?

Again, I congratulate the authors on a very interesting paper and particularly for having such well organized records of these unusual cases, even after 25 years.

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DR EDWARD L. RAAB. In cases of closed head trauma, even when there has been a sixth nerve palsy, and also in cases of diabetes, it would be wise to check accommodative amplitudes. In both these conditions the ability to accommodate can be reduced. I have diagnosed unknown diabetes in patients with accommodative insufficiency. This is another type of accommodative esotropia, which D. Costenbader called the accommodative effort syndrome. It's another way in which the supply of accommodation does not match the demand. Supplying the appropriate refractive correction will benefit these patients. Therefore this is an additional useful diagnostic test in these cases.

DR MALCOLM L. MAZOW. I would like to complement Dr Pollard on his own mental electronic records. It would be interesting to look at the sensory mechanism that is present in these older individuals who have had some sort of trauma. Over the years in medical legal situations where a child has developed strabismus, I have found the problem is often a breakdown of a monofixational syndrome. When you realign the eyes of these children and they appear to be straight, they have at best 60 to 70 seconds of arc of stereopsis, and they do have suppression by the 4 base out prism test or a 20 base in test.

DR GUNTER K. VON NOORDEN. I was most intrigued by the authors' observation of highly myopic and esotropic children who, after correction of their myopia became orthotropic. In my career as a strabismologist. I have never encountered this clinical event which the authors classified together with esotropia secondary to an uncorrected hypermetropia as "accommodative strabismus." I don't doubt for a moment the authors' observation but

cannot agree with this classification since any accommodative effort in these children would increase the myopia and thus further decrease the visual acuity. I would like to ask whether Dr Pollard has any other explanation for this phenomenon which must be extraordinary rare indeed.

DR ZANE F. POLLARD. Dr Raab, I did not check accommodative ability in these patients, but this is a point worth taking forward in patients we will see in the future.

Dr Mazow, the older patients, these were not monofixators. While the purpose of the paper was to present unusual cases of accommodative esotropia, I did not include all of the sensory status in these patients. The trauma patients were all bifixators.

Dr Von Noorden, I do not have the answer as to what is going on in these little kids with the myopia. I have only seen 2 in 25 years, so that is not a tremendous number. We are talking about 2 patients less than 6 months with high myopia and accommodative esotropia. One broke down and eventually did require surgery. All I can say is that this child must have been over accommodating because when we gave him the glasses, his eyes became straight; both of them did become straight in their glasses. One has remained straight until this time. He is now a teen-ager. So we have a 14-year follow-up and he does have a small esophoria without his glasses.

Dr Stager, referring to the article by Birch, Ken Wright and Stager, most of their patients probably do represent a different sub-segment of patients. Most of their patients, which did require surgery, had more than 40 prism diopters of hyperopia. Most of our patients had more than 3.5 diopters of hyperopia and were less than 45 prism diopters of esotrpia. These patients obviously do represent a different subsegment because none of these patients had DVD and none of them had latent nystagmus both of which we associate with congenital esotropia. In terms of sensorial status can we use the stereopsis development as distinguising these 2 groups? I don't know. I originally thought yes until Ken Wright came out with his series of children age 13-19 weeks that he had operated on with good stereopsis results. I think even one of his patients had 40 seconds. Several others also had high levels of stereopsis. These children in my series are now older and I have now measured them. I do not have any in this group with the onset of accommodative esotropia less than 6 months of age with 40 seconds of stereopsis. The best was 60 seconds; most were in the 140 to 400 seconds of stereopsis range. The 2, which did end up having surgery, did not have high grades of stereopsis but were in the 80 to 400 seconds range.

In terms of motion asymmetry, what Dr Stager was referring to was taking children and covering one eye and rotate the OKN drum nasal to temporal and then temporal to nasal. Children with congenital esotropia, as shown by Ken Wright, have a strong temporal to nasal bias; the OKN nystagmus is much brisker going temporal to nasal compared with rotating the drum nasal to temporal. Children that are normal and children with accommodative esotropia do not have this bias. I did not report on this, but I did try to check this in some of these young patients. I found it extremely difficult to perform this test in children 2-4 months of age. Also Ken Wright has shown that a lot of children in the first few months of life turn out to be normal and don't develop esotropia, can have this temporal nasal bias in the first few months of life. This usually dissipates by 4 months of age. What I will do and what I feel is a good point, (and you have helped me to better understand this group of patients) now that these patients are older, is do the OKN testing to see if these patients have the temporal to nasal bias as is typical of congenital esotropia or will they look more like a regular accommodative esotropia with normal OKN nasally and temporally.

I want to thank Dr Stager again for his comments and the rest of the discussants. I want to thank Dr Jones and the program committee for allowing me to present my paper.