Acute Acquired Concomitant Esotropia
Clinical features, Classification, and Etiology

Jingchang Chen, MD, PhD, Daming Deng, MD, Yuan Sun, MD, PhD, Tao Shen, MD, PhD, Guobin Cao, MD, Jianhua Yan, MD, PhD, Qiwen Chen, MB, and Xuelian Ye, MB

Abstract: Acute acquired concomitant esotropia (AACE) is a rare, distinct subtype of esotropia. The purpose of this retrospective study was to describe the clinical characteristics and discuss the classification and etiology of AACE.

Charts from 47 patients with AACE referred to our institute between October 2010 and November 2014 were reviewed. All participants underwent a complete medical history, ophthalmologic and orthoptic examinations, and brain and orbital imaging.

Mean age at onset was 26.6 ± 12.2 years. Of the 18 cases with deviations ≤20 PD, 16 presented with diplopia at distance and fusion at near vision at the onset of deviation; differences between distance and near deviations were <8 PD; all cases except one were treated with prism and dioptria resolved. Of the 29 cases with deviations >20 PD, 5 were mild hypermetropic with age at onset between 5 and 19 years, 16 were myopic, and 8 were emmetropic with age at onset >12 years; 24 were surgically treated and 5 cases remained under observation; all 24 cases achieved normal retinal correspondence or fusion or stereopsis on postoperative day 1 in synoptophore; in 23 cases diplopia or visual confusion resolved postoperatively. Of the 47 cases, brain and orbital imaging in 2 cases revealed a tumor in the cerebellopontine angle and 1 case involved spinocerebellar ataxia as revealed by genetic testing.

AACE in this study was characterized by a sudden onset of concomitant nonaccommodative esotropia with diplopia or visual confusion at 5 years of age or older and the potential for normal binocular vision. We suggest that AACE can be divided into 2 subgroups consisting of patients with relatively small versus large angle deviations. Coexisting or underlying neurological diseases were infrequent in AACE.

INTRODUCTION

A acute acquired concomitant esotropia (AACE) represents a relatively rare, distinct subtype of esotropia, which occurs in older children, adults, and even the elderly.1–4 It was reported that AACE made up 0.3% of childhood strabismus.5 AACE is characterized by a sudden onset of concomitant esotropia with diplopia.1–3,6,7 In 1958, Burian and Miller reported 5 cases of AACE and described the 3 categorical features of AACE, which have been accepted as the fundamental symptoms and the pivotal work in this area.1 Since then, findings from a number of reports have added to this work.6–8–10 However, cases series in most of these reports were quite limited, and, there remain disputes over its etiology, clinical features, and classification. In this report, we present 47 cases with AACE, describe its clinical characteristics and management, and discuss its classification as well as its relationship with neurologic diseases.

PATIENTS AND METHODS

Charts of patients diagnosed with AACE were reviewed. These records were derived from cases seen from October 2010 to November 2014 at the Pediatric Ophthalmology and Strabismus Services of the Zhongshan Ophthalmic Center of Sun Yat-Sen University. This study adhered to the tenets of the Declaration of Helsinki and was approved by the ethics committee of the Zhongshan Ophthalmic Center of Sun Yat-Sen University (Guangzhou, China).

Patients who met the following criteria were included in this study: (1) concomitant esotropia with acute onset (deviation in all directions of gaze differing by ≥2 prism diopters) when primary deviations were ≤20 prism diopters and by ≤5 prism diopters when primary deviations were >20 prism diopters), (2) patients experiencing diplopia or visual confusion, and (3) best corrected visual acuity of not <0.5 in each eye. Patients with a history of eye surgery or those with a reduction of ≥10 PD in their esotropia with a full hyperopic spectacle correction for 1 month were excluded.

Comprehensive medical histories, ophthalmologic, and orthoptic examinations were performed on all patients. The age at onset was determined by the ophthalmologist according to the medical history and patients’ photographs. Deviation size was assessed using prism and alternate cover tests in the cardinal position gazes with refractive correction. Synoptophore was used to evaluate the 3 grades of binocular single vision. Cycloplegic refraction was performed after administration of...
1% atropine ointment once daily for 3 days in patients under 7 years old and 1% cyclopentolate eye drops every 5 min for 3 times for those >7 years old. A full cycloplegic refraction was prescribed for those with refractive errors ≥1.0 diopter (D) (spherical equivalent). Additionally, all patients received brain and orbital computed tomography scans, and, in some cases, magnetic resonance imaging. Neurological examinations were performed and comprehensive medical histories were taken for cases with intermittent esotropia or variable deviations to rule out cyclic esotropia and myasthenia gravis.

**RESULTS**

A total of 47 cases were included in this study, with 18 cases showing deviations ≤20 PD (Figure 1) and 29 with deviations >20 PD (Figure 2). There were 25 men and 22 women. Age at onset was 26.6 ± 12.2 years (range: 5.0–59.0 years). The follow-up period was 1.8 ± 0.9 years (range: 0.5–4.0 years). Slitlamp and funduscopy examinations revealed no abnormality. Mild hypermetropia was observed in 5 cases, 9 cases were emmetropic, and 33 had myopia. Cerebellopontine angle tumors were present in 2 cases and 1 patient experienced spinocerebellar ataxia. All cases showed normal retinal correspondence or fusion or stereopsis in the synoptophore before therapy. Diplopia or visual confusion resolved in 40 cases following prism or surgical treatment.

Clinical features of cases with deviations ≤20 PD are summarized in Table 1. Of these 18 cases, none had a family history of strabismus; 1 case resulted following 24 h of driving; 1 case who presented with esodeviation accompanied by nystagmus and poor coordination of gait and was subsequently diagnosed as spinocerebellar ataxia as revealed from genetic testing; 1 case had a history of chemotherapy and radiotherapy for nasopharyngeal cancer 8 years prior; 14 cases had diplopia at distance but single vision at near vision; 2 cases initially had distance diplopia and later developed near diplopia; 2 cases had both distance and near diplopia at onset; 17 cases were myopic (−0.75 to −9.75D) and 1 case was emmetropic; 15 cases with myopia were using refractive corrections at presentation; in the synoptophore, all 18 cases showed preoperative normal retinal correspondence and stereopsis; brain and orbital imaging were not remarkable; the duration between onset and treatment ranged from 2 weeks to 10 years; diplopia resolved after being corrected with bilateral base out prisms of 2 to 7 PD in all but the one case of spinocerebellar ataxia.

Clinical features of cases with deviations >20 PD are summarized in Table 2. Of these 29 cases, none had a family history of strabismus; brain and orbital imaging in 2 cases revealed masses in the cerebellopontine angle which consisted a 15 × 11 mm lipoma or a 40 × 23 mm cholesteatoma; 5 cases involved mild hypermetropic (+1.0 to +1.75D) with age at onset between 5 and 19 years, 16 cases were myopic (−0.75 to −11.75D) with age at onset between 15 and 59 years, and 8 cases were emmetropic with age at onset between 12 and 38 years; 3 cases experienced visual confusion, 1 case had diplopia at distance whereas both distance and near diplopia were present in the others; 4 cases showed an occasional orthophoria; 1 case whose brain imaging revealed cholesteatoma showed bilateral nystagmus; in the synoptophore, 29 cases showed preoperative normal retinal correspondence or fusion or stereopsis; 4 cases had a minimal (<1) bilateral lateral rectus muscle under-actions; 16 cases were subjected to unilateral medial rectus recession and lateral rectus resection, 7 cases received bilateral or unilateral medial rectus recession, 1 case involved a unilateral lateral rectus resection, 5 cases remained under observation; age at surgery was 26.0 ± 2.7 years (range: 5.5–64.0

**FIGURE 1.** Composite 9-gaze photograph of an AACE patient with a relatively small angle. A 17-year-old boy with distance diplopia for 2 years, presented 20 PD of concomitant esotropia, normal ocular version and duction, normal stereopsis in synoptophore, and a myopia of 2.25 D. His diplopia resolved following treatment with bilateral base out prisms of 7 PD. AACE = acute acquired concomitant esotropia.
years); the duration between onset and surgery was 2.4 ± 0.3 years (range: 0.5–5.0 years); the interval between diagnosis and surgery was 0.4 ± 0.1 years (range: 0.1–1.0 years); in 23 cases diplopia or visual confusion resolved at 1 day to 2 months postoperatively and diplopia remained in 1 case for residual deviation; all 24 cases achieved normal retinal correspondence or fusion or stereopsis on postoperative day one in synoptophore; with the exception of 1 case, all 24 cases were aligned within 10 PD or less of orthotropia; no recurrences were observed at postoperative follow-up as performed at 0.5 to 3 years; no improvements were observed for the patients under observation.

DISCUSSION

Acute acquired concomitant esotropia (AACE), or acute late-onset concomitant esotropia, are generally agreed to be characterized by a sudden onset of concomitant esotropia with diplopia after infancy or in older children and adults.1–4,6 Some reports describe AACE as being an acute-onset condition confined to children ≥5 years of age, which might be one of the primary features that differentiates it from acquired non-accommodative esotropia.2,14 However, in several other studies it was reported that the acute deviation developed during infancy and early childhood7,8 and diplopia was an inconsistent feature.4,8 Here, we reviewed a large case series of AACE, and, the salient characteristics observed were: (1) concomitant non-accommodative esotropia with acute onset, (2) an accompanying diplopia or visual confusion, (3) potential for normal binocular vision and highly favorable surgical outcomes for those with deviations >20 PD, (4) ≥5 years old at the time of onset, and (5) no tendency for spontaneous resolution.

Burian and Miller are generally considered as having performed the initial pivotal work in the field of AACE.1

TABLE 1. Clinical Features of Cases With Deviations ≤ 20 PD (n = 18)

| Age Onset (Year) | Refraction (Spherical Equivalent) | Fusion(4) Before Corrected With Prism | Deviation (PD) |
|------------------|-----------------------------------|--------------------------------------|----------------|
|                  | RE                                | LE                                  |                |
| Average          | 32.3 ± 10.8                       | −3.5 ± 2.3                          | 9.6 ± 6.3      | 9.3 ± 4.4 |
| Range            | 16.0 to 51.0                      | 0 to −9.5                           | 6.1 ± 1.6      | 11.3 ± 3.9 |
|                  | 16.0 to 51.0                      | −0.25 to −9.75                     | 4.0 to 28.0    | 3.0 to 20.0 |
|                  |                                   |                                     | 2.0 to 10.0    | 5.0 to 20.0 |

LE = left eye, PD = prism diopter, RE = right eye.
TABLE 2. Clinical Features of Cases With Deviations >20 PD (n = 29)

| Age Onset (Year) | Refraction (Spherical Equivalent) | Corrected Visual Acuity | Deviation (PD) |
|------------------|-----------------------------------|-------------------------|----------------|
|                  | RE | LE | RE | LE | Near | Distance |
| Average          | 23.1 ± 11.9 | −2.1 ± 3.1 | −2.0 ± 3.1 | 1.0 ± 0.2 | 1.0 ± 0.2 | 41.0 ± 13.5 | 40.7 ± 14.0 |
| Range            | 5.0 to 59.0 | +1.75 to −11.0 | +1.75 to −10.0 | 0.5 to 1.5 | 0.6 to 1.5 | 25.0 to 70.0 | 20.0 to 70.0 |

LE = left eye, PD = prism diopter, RE = right eye.

According to their findings, AACE can be classified into 3 types: (1) the Swan type, which results from temporary or permanent monocular occlusion or visual loss, (2) the Franceschetti type, which is associated with low hyperopia, a minimal accommodative element and relatively large angle of deviation, and (3) the Bielschowsky type, also be classified as divergence paralysis, which occurs in patients with myopia (≤ 5 diopters) following physical or psychic shock or exhaustion and exhibits a diplopia for distance but fusion for near vision. Bielschowsky claimed that uncorrected myopia played a role in the etiology of this form of strabismus. In our study, none of the cases experienced monocular patching or visual loss, thus the criteria for a Swan type was not achieved in any of these cases. As the suspension of normal fusion was the underlying cause of sensory strabismus, we propose that the Swan type of AACE should be integrated into a category of sensory esotropia. Moreover, of these 18 cases the etiology of AACE remains unclear, it is important to emphasize the infrequent, but possible, presence of an underlying or coexisting neurological condition in patients with AACE. This would be a particular concern for those who present with other neurologic signs or accompanying nystagmus. Consistent with previous studies,12–14 we suggest that a neurological and/or neuroradiological investigation are warranted for AACE under conditions where neurological findings (such as headache, papilloedema, clumsiness, poor motor coordination, etc.), or nystagmus are present.

The etiology of AACE is elusive. In our study, most of the cases had normal corrected visual acuity, binocular vision potential and presented no other pathologic signs other than the esotropia. Therefore, we do not deem uncorrected myopia, accommodative element, physical or psychic exhaustion, decompensated monofixation syndrome or neurologic disorders as the genesis of AACE. However, coexisting or underlying neurological pathologies were present in 6.4% of the AACE cases in this study. Several reports have described patients with intracranial diseases who present with, or as, acute concomitant esotropia.10–13 In the report of Liu GT, 40% of children with acquired esodeviation associated with an identifiable neurologic insult showed concomitant esodeviation.15 Such findings support the conclusion that neurologic disease may cause acquired concomitant esotropia, though for most cases with concomitant esotropia its cause is benign. In addition, in our study all 3 cases with neurological pathologies were associated with cerebellar diseases and 2 presented with nystagmus. These results suggest that the cerebellum, as a part of extrapyramidal system, may play an important role in maintaining normal ocular alignment. Although the genesis of AACE remains unclear, it is important to emphasize the infrequent, but possible, presence of an underlying or coexisting neurological condition in patients with AACE. This would be a particular concern for those who present with other neurologic signs or accompanying nystagmus. Consistent with previous studies,10,12 we suggest that a neurological and/or neuroradiological investigation are warranted for AACE under conditions where neurological findings (such as headache, papilloedema, clumsiness, poor motor coordination, etc.), or nystagmus are present.

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