Reprinted from the *Journal of Pediatric Ophthalmology & Strabismus* 1987; 24(4):176-177.

Adie's Syndrome as a Cause of Amblyopia

A.M. Agbeja, M.D.
G.N. Dutton, M.D.
Glasgow, Scotland

ABSTRACT

Adie's syndrome comprises a tonic pupil, which may be associated with impairment of accommodation, in the presence of diminished or absent deep tendon reflexes. We report a case of a 4-year-old boy with Adie's syndrome in which latent hypermetropia was made manifest by accommodative paresis and resulted in reversible amblyopia.

Introduction

Adie's syndrome is known to affect patients of either sex and of any age, with an unexplained predilection for women in the third to fifth decades.[1-3]

Adie's syndrome is rare in children. Adie's youngest patient was 17 years old and the youngest case in Holmes' series was 10 years of age. Esterly et al[4] described bilateral tonic pupils in a 3-year-old girl with autonomic dysfunction and Goldberg[5] described the condition in 15 children with familial dysautonomia (the Riley-Day syndrome). In none of these cases was amblyopia reported.

We report a case of Adie's syndrome in which impairment of accommodation was the probable cause of amblyopia.

Case Report

An intelligent and cooperative 4-year-old boy was first seen in our clinic in March 1986. Two of his school friends had told him, in December 1985, that "one eye was bigger than the other," and his mother subsequently noticed that the right pupil was larger than the left. A pre-school eye test in March 1985 had revealed no impairment of visual acuity. There was no history of antecedent viral illness. At presentation, his visual acuities were 6/36, N24 (0.12) right eye and 6/6, N5 (0.6) left eye without correction, and with a pinhole. His right pupil was enlarged and reacted slowly to accommodation but not to light, the left pupillary reactions were normal. The near point of accommodation was 20 cms for the right eye and 6 cms for the left eye.

FIGURE: The appearances of the pupils before (A) and after (B) the instillation of pilocarpine 0.125%.

Fifteen minutes following instillation of pilocarpine 0.125%, the right pupil constricted from 6 mm to 2 mm but the left pupil remained at 3 mm in diameter (Figure).

The biceps jerks were absent and the knee jerks were sluggish but improved on reinforcement. No other neurological signs were elicited.

Cycloplegic refraction revealed bilateral hypermetropia of +1.50DS/+0.50DC axis 90 right and +0.50DS left. A spectacle correction was prescribed with an executive bifocal segment for the right eye. With this correction the visual acuity was found to be 6/18 in the right eye and 6/6 in the left. A therapeutic trial of partial occlusion of the left eye was then commenced. After 2 weeks of patching and constant wearing of spectacles, the visual acuity of the right

From the Tennent Institute of Ophthalmology, Western Infirmary, University of Glasgow, Glasgow, Scotland.

Requests for reprints should be addressed to G.N. Dutton, M.D., Tennent Institute of Ophthalmology, Western Infirmary, University of Glasgow, Glasgow G11 6NT, Scotland.

eye had improved to 6/6, N5 with glasses and the acuity of the left remained unchanged at 6/6, N5. His visual acuities have remained the same on subsequent follow-up. Further evaluation to elicit evidence of spontaneous recovery of accommodation is currently being carried out.

Discussion

To our knowledge, Adie's syndrome is rare in childhood and has not previously been reported as a cause of amblyopia.

Adie's syndrome is characterized by a tonic pupil which may be associated with hypoactive or absent deep tendon reflexes. It is a common anomaly which runs a benign course and, in the absence of other signs or symptoms, heralds no neurological or systemic disease state.[6]

The tonic pupil was first described by Markus in 1906,[7] and later by Adie in 1931,[6] who called it the Pseudo-Argyll Robertson pupil. Adie described five patients whose pupils did not react to light but reacted tonically to accommodation and whose deep tendon reflexes were diminished to varying degrees. In the same year, Holmes[8] described 54 patients with tonic pupils, 19 of whom demonstrated other neurological anomalies, chiefly loss or diminution of the deep tendon reflexes. The same combination of clinical signs had also been described previously by Parkes Weber in 1923.[9]

Non-specific viral illness, herpes zoster ophthalmicus, varicella,[10] orbital injuries,[11] dysautonomia,[5] and bronchial carcinoma associated with the myasthenic syndrome[12] have all been rarely implicated as causes of Adie's syndrome; however, in the majority of cases, the etiology is indeterminable.

The pupillary features of Adie's syndrome include: a) relative mydriasis in bright illumination; b) a poor to absent reaction of the pupil to light; c) slow pupillary contraction with prolonged near effort; d) slow redilation after near effort; e) sectional paralysis of the iris associated with vermiform movements of the pupillary margin[13]; f) accommodative paresis[14]; and, g) supersensitivity to pilocarpine 0.125% or mecholyl 2.5%.[1,15,16]

Post-mortem studies have revealed degeneration of the ciliary ganglion on the affected side associated with partial atrophy of the sphincter pupillae and minimal neuronal degeneration of the sacral dorsal root ganglion, the superior cervical ganglion and the sciatic nerve. No underlying cause for these abnormalities has been found.[11,17,18]

We presume that in this case, latent hypermetropia was "unmasked" by acquired accommodative paresis. This resulted in impairment of visual function for both distance and near with subsequent reversible amblyopia.

A child presenting with significant impairment in visual function associated with a dilated, poorly reacting pupil, provides cause for concern, and an awareness that Adie's syndrome may give rise to these features may diminish the need for extensive neurological investigation in the first instance.

Adult patients with Adie's syndrome may also present with blurring of vision in one eye due to accommodative paresis combined with hypermetropia or presbyopia.[19] Although this is commonly a self-limiting problem, appropriate spectacle correction can provide considerable symptomatic improvement for these patients.[7]

Acknowledgments

We thank Dr. J.B.P. Stephenson for providing a detailed pediatric neurological assessment, and Alison Pike for providing orthoptic assessment and follow-up.

References

1. Loewenfeld IE, Thompson HS: The tonic pupil: A re-evaluation. *Am J Ophthalmol* 1967; 63:46-87.
2. Talbot MD: The Holmes-Adie syndrome — A report of three cases and observations on its natural history. *Br J Clin Pract* 1981; 35:163-166.
3. Press LJ: Management of Adie syndrome. *Am J Optom Physiol Opt* 1981; 58:235-238.
4. Esterly NB, Cantolino SJ, Alter BP, et al: Pupillotonia, hyporeflexia, and segmental hypohidrosis: Autonomic dysfunction in a child. *J Pediatr* 1968; 73:852-859.
5. Goldberg MF, Payne JW, Brunt PW: Ophthalmologic studies of familial dysautomonia: The Riley-Day syndrome. *Arch Ophthalmol* 1968; 80:732-743.
6. Adie WJ: Pseudo-Argyll Robertson pupils with absent tendon reflexes: A benign disorder simulating tabes dorsalis. *Br Med J* 1931; 1:928-930.
7. Markus C: A peculiar pupil phenomenon in cases of partial iridoplegia. *Trans Ophthalmol Soc UK* 1906; 26:50-58.
8. Holmes G: Partial iridoplegia associated with symptoms of other diseases of the nervous system. *Trans Ophthalmol Soc UK* 1931; 51:209-224.
9. Parkes Weber F: Dr. Markus' original case of Markus' syndrome shown twenty seven and a half years ago. *Proc R Soc Med* 1933; 26:530-535.
10. Duke-Elder S, Scott GI: *A System of Ophthalmology.* London, Henry Kimpton, 1971, vol 12, pp 681-688.
11. Harriman DGF, Garland H: The pathology of Adie's syndrome. *Brain* 1968; 91:401-418.
12. Walsh FB, Hoyt WF: *Clinical Neuro-ophthalmology,* ed 3. Baltimore, Williams & Wilkins, 1986.
13. Thompson HS: Segmental palsy of the iris sphincter in Adie's syndrome. *Arch Ophthalmol* 1978; 96:1615-1620.
14. Bell RA, Thompson HS: Ciliary muscle dysfunction in Adie's syndrome. *Arch Ophthalmol* 1978; 96:638-642.
15. Pilley SFJ, Thompson HS: Cholinergic supersensitivity in Adie's syndrome: Pilocarpine vs Mecholyl. *Am J Ophthalmol* 1975; 80:955.
16. Bourgon P, Pilley SFJ, Thompson HS: Cholinergic supersensitivity of the iris sphincter in Adie's tonic pupil. *Am J Ophthalmol* 1978; 85:373-377.
17. Harriman DGF: Pathological aspects of Adie's syndrome. *Adv Ophthalmol* 1970; 23:55-73.
18. Ruttner F: Die tonische pupillenreaktion. Klinische und anatomische untersuchungen. *Mschr Psychiat Neurol* 1947; 114:265-330.
19. Scheinberg IH, Adler RI: Adie's syndrome: Case report. *Ann Ophthalmol* 1979; 11:247-248.