Long term prognosis of transient lone bilateral blindness in adolescents and young adults

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Abstract
We describe a group of 14 patients aged 8-38 years at presentation who had one or more sudden transient attacks of bilateral blindness. Eight patients described bilateral blindness as their only symptom whereas six others experienced some mild associated symptoms. Visual loss always developed within seconds and attacks were often precipitated by exercise, stress, or postural change. Of 13 patients available for review, none suffered a major vascular event during a mean follow up of 10 years. When adolescents and young adults present with transient bilateral blindness, investigations are unlikely to reveal a cause and the long term prognosis appears benign.

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Transitum bilateral blindness can occur as a result of vertebrobasilar ischaemia, migraine, various angiographic procedures, and during epileptic seizures. When sudden transient bilateral blindness occurs in the elderly as the only symptom, there is debate as to whether the cause is migraine or a transient ischaemic attack. Although bilateral visual loss may be part of the syndrome of basilar artery migraine, there are usually accompanying symptoms such as ataxia, dysarthria, vertigo, tinnitus, bilateral peripheral dysesthesiae, disturbance of consciousness, and occipital headache. However, when transient bilateral blindness is the only symptom, and onset is sudden, a diagnosis of migraine is much less tenable; a recent report from the Oxfordshire Community Stroke Project came to the conclusion that the cause was cerebral ischaemia, consistent with earlier reports of vertebrobasilar occlusive disease. By contrast, similar symptoms in adolescents and young adults seem most unlikely to be ischaemic in origin, at least in the sense of being caused by embolism from the heart or arterial disease. As, over the years, we have seen several such patients, we thought it would be helpful to describe their clinical features and, more importantly, their long term prognosis.

Methods
To define the long term prognosis of transient lone bilateral blindness in adolescents and young adults, we reviewed the case records of 14 patients who had experienced one or more attacks and who were referred to neurologists in Oxford and Sheffield. The cases are probably not consecutive and presumably other patients would never have been referred to us by their general practitioner or ophthalmologist. With the exception of one patient, they were first seen between 1978 and 1983. We have previously defined lone bilateral blindness as the sudden onset of dimming or loss of vision over both visual fields simultaneously, which lasted less than 24 hours and had no associated symptoms of focal cerebral dysfunction, epilepsy, or change in consciousness. In this paper we have restricted the term to refer to patients with complete blindness and with no positive visual phenomena, such as flashing lights, suggestive of migraine. Eight patients satisfied this strict definition but, in the remaining six, there were some accompanying symptoms. In 1991 we tried to follow up all 14 patients using a postal and/or telephone questionnaire to ascertain the continuing frequency, duration, precipitants, and associated features of the attacks. We also questioned the patients verbally or by questionnaire about symptoms of possible transient ischaemic attacks, stroke, other vascular events, and epilepsy. The duration of follow up was from the date when first assessed by the neurologist to the completion of the questionnaire.

Patients
The following eight cases are examples of lone bilateral blindness.

CASE 1
An 8 year old girl presented with a six month history of frequent attacks of visual disturbance in which the whole of her visual field became momentarily either black or white, several times a day, and without obvious precipitants. There was a single episode of total loss of vision lasting two minutes during which she fell off a chair because she could not see. She had numerous other episodes of flashing throughout the visual fields of both eyes but none of the attacks was associated with headache. Distinct from the visual symptoms, she had two episodes of migraine headache, each lasting several days, but without visual or gastrointestinal disturbance. Apart from hyperextensible joints, clinical
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examination was normal. A full blood count, routine biochemistry, radiographs of skull, orbits and chest, and CT brain scan were normal. The attacks pattered out after one year and she remained well up to nine years after her initial presentation.

CASE 2
A 16 year old schoolgirl gave a five year history of brief visual blackouts; there were very few days when she was free of attacks. She described the sudden onset of blindness as coming from one or other side, lasting only seconds, with a gradual fluctuating recovery, but no headache. Sensorineural deafness had been noted when she was 13 years old. She had a sister who was epileptic. Apart from deafness, neurological examination was normal. A CT brain scan, lumbar puncture, and visual evoked responses were normal but an EEG showed bilateral sharp waves with excess slow components. She continues to have attacks precipitated by stress after nine years of follow up.

CASE 3
A 16 year old schoolgirl described an episode when she was cycling uphill, during which she felt rather weak and then, over a few seconds, lost vision bilaterally. Her vision went completely for about 60 seconds and gradually returned. There was no associated headache or past history of headache. Her brother had migraine. Clinical examination and visual evoked responses were normal. Over the next 10 years she continued to suffer attacks, about 40 in total, usually with exertion and only ever when standing. During some of these she felt unsteady and appeared pale, but she never felt faint or lost consciousness. Otherwise she remained well.

CASE 4
A 17 year old trainee timber assembler described his first attack of visual loss at the age of nine when, without warning, he suddenly became blind and was unable to see for 10 minutes. There were no associated features but he continued to experience attacks lasting for five minutes to one hour twice weekly. They were usually related to activity or standing and several attacks occurred 15 minutes after using a "Bull-worker" exercise machine. On three occasions he described slight weakness of his left arm and both legs for several minutes, but only after the attack of blindness had resolved. He had smoked 40 cigarettes daily since the age of nine. His father, grandfather, great-grandfather and paternal uncle all suffered migraine affecting their vision. Neurological examination was normal. Loud subclavian bruits were present but varied with posture. The blood pressure was normal and equal in both arms. The brachial pulses could be obliterated by raising his arms above his head. The patient declined an arch aortogram to exclude subclavian stenosis. A full blood count, routine biochemistry, chest and cervical spine radiographs, echocardiography, electrocardiography, and 24 hour ECG were normal. He did not reply to a questionnaire but, according to his GP, he was well 10 years later.

CASE 5
An 18 year old boy gave a two year history of weekly episodes of blindness one month following a head injury when he lost consciousness for about one minute. They began with central black spots spreading bilaterally within seconds to involve all but the periphery, lasting from one to five minutes and never followed by headache. Central vision was often so black that he had walked into doors or fallen downstairs. Some of the attacks were brought on by getting up from stooping or by walking. His mother suffered from migraine without aura. Clinical examination, resting ECG, and EEG were normal. The attacks stopped two months after presentation and he was well 11 years later.

CASE 6
A 20 year old RAF ground crew airman suddenly became bilaterally blind late one night after drinking with friends. An hour later he complained of a mild, left sided, throbbing headache, which lasted only 15 minutes; he vomited one hour later. There was no past or family history of migraine. Neurological and general physical examination were unremarkable except for almost total blindness in both eyes with only slight appreciation of a bright torch light, slightly better on the left than the right. There was only a faint blink to menace. Recovery started within three hours, first with a right homonymous hemianopia, and was complete within four hours. Unfortunately he was lost to follow up.

CASE 7
This 34 year old man was driving home from work when quite suddenly he lost all his vision. He could see nothing at all for a second or two, then quite suddenly his vision returned, "like a light that had been switched off and then on again". There were no associated symptoms, although over previous days he had noted episodic pulsation in the centre of his forehead, and may have had a similar feeling just before he lost his vision. For many years he had had attacks of migraine with visual aura, quite different from the present episode, which was not accompanied by headache or vomiting. Clinical examination was normal and he only ever had the single episode. Twelve years later he was well.

CASE 8
A 38 year old woman was walking when quite suddenly her vision went completely black in both eyes for a matter of seconds. This blindness was then replaced by a grey haze through which she could see faint outlines; gradually this improved to normal in about an hour. A similar episode occurred six weeks later while standing washing up, but it was two hours before her vision fully recovered. "My sight just went completely, I could see nothing although I knew where I was and was aware
of everything being said around me." A third and final episode occurred nine months later. There was a family history of migraine. Clinical examination, full blood count, routine biochemistry, and ECG were normal. Eight years later she was well.

Six other cases, three of each sex and aged 12–36 years at first presentation, described some other mild and transient symptoms during their attacks of bilateral blindness. These included dizziness, unsteadiness, light-headedness, vertigo, sweating, pallor, and pins and needles in one arm. Investigations among these patients included ECGs, 24-hour ECGs in two, echocardiograms in three, CT scans in two, EEGs in one, and cerebral angiography in two. These failed to demonstrate a definite cause in any patient.

Follow up

Twelve patients completed the questionnaire. After a mean of 10 years (range 4–13), all patients were alive; only two continued to have attacks of blindness (cases 2 and 3) and no patient had a permanent visual deficit, stroke, myocardial infarction, or epilepsy. One other patient (case 4) was alive and asymptomatic according to his local doctor and one (case 6) was lost to follow up.

Discussion

We have described a group of adolescents and young adults who presented with sudden attacks of transient bilateral blindness. We were unable to demonstrate a definite cause or mechanism for the blindness in any of these patients, in spite of quite extensive investigation in some. In the patients with associated, although mild, symptoms, presyncope seemed a likely mechanism although none had episodes of true syncope, even on other occasions, and one patient had attacks of blindness while supine. In one patient with associated pins and needles hyperventilation seemed a possible cause. The question of migraine arises, but the blindness developed within seconds, not minutes, and there were no positive visual phenomena at presentation. Only one patient described separate episodes in which she experienced either flashing lights or headache. Headache occurred in one other patient during an episode. However, in favour of a migrainous aetiology, there was a past or family history of migraine in five of the eight patients who had no associated symptoms. The sudden onset of these episodes raises the possibility that they are epileptic phenomena, although true epileptic blindness is rare and is usually pre- or postictal. Attacks of complete blindness with no other seizure activity, confirmed by bi-occipital EEG spike-wave activity on photic stimulation, have been described, 1 but none of our patients developed epilepsy during follow up. Transient ischaemic attacks affecting the occipital cortex or both retinæ simultaneously could cause lone bilateral blindness, 11 but this seems unlikely given the patients’ ages, lack of vascular risk factors, and benign prognosis. The frequent, but not invariable, precipitation of the attacks by exercise and postural change suggests that a haemodynamic challenge might be required. 13 Transient monocular blindness has been similarly reported after bending or exercising. 14

Although we can speculate, in truth the cause of the attacks in our patients is unknown. We have described a group of adolescents and young adults who have sudden and brief attacks of complete bilateral blindness with no other symptoms of focal neurological dysfunction. In some, attacks may be precipitated by exercise, stress or postural change, but the mechanism of visual loss is unclear and investigations are unlikely to reveal any relevant abnormalities. Unlike older patients with lone bilateral blindness, who have a significant risk of subsequent stroke, transient bilateral blindness in adolescents and young adults is usually self-limiting and the long-term prognosis is excellent. Such patients should not be subjected to potentially hazardous diagnostic procedures and can be reassured that these frightening attacks are benign.

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