



## Postgraduate Medicine

ISSN: 0032-5481 (Print) 1941-9260 (Online) Journal homepage: http://www.tandfonline.com/loi/ipgm20

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### Walter G. Drymalski

To cite this article: Walter G. Drymalski (1980) Cortical blindness, Postgraduate Medicine, 67:4, 149-156, DOI: 10.1080/00325481.1980.11715430

To link to this article: http://dx.doi.org/10.1080/00325481.1980.11715430



Published online: 07 Jul 2016.

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# **Cortical blindness**

The changing incidence and shifting etiology

Walter G. Drymalski, MD

## Consider

What are the most common characteristics of cortical blindness?

Under what circumstances is the disorder difficult to diagnose?

Which diagnostic test is the most useful for follow-up evaluation?

The incidence and etiology of cortical blindness—a disorder that usually occurs after cerebrovascular accidents, cardiorespiratory arrest, severe head trauma, or bacterial meningitis—may be changing with the development of effective means of cardiopulmonary resuscitation, as three cases discussed here show.

For a long time cortical blindness has been considered a rare event resulting from bilateral damage to the occipital lobes or the optic radiations of the brain beyond the lateral geniculate body. The causes are few.

In the pediatric age-group, causes include head trauma, meningitis, and cardiorespiratory arrest.<sup>1-3</sup> The head trauma is usually sufficiently severe to result in generalized CNS depression, but prognosis for recovery of sight is favorable.<sup>2</sup> Transient cortical blindness in children after seemingly minor head trauma also has been reported.<sup>1</sup> Cortical blindness as a sequela of bacterial meningitis is rare, and the incidence attributable to each of the various pathogens is not known.

In adults, the most common causes are cerebrovascular accidents and episodes of cardiorespiratory arrest. In the largest series to date (58 cases), Symonds and Mackenzie<sup>4</sup> reported in 1957 that cortical blindness was caused almost exclusively by thrombosis or embolization of cerebral vessels. They believed that most of the episodes of cortical blindness were caused by microemboli from vertebral or basilar artery plaques or thrombi lodged in the distal posterior circulation. In a smaller series reported in 1957 by Bergman,<sup>5</sup> cerebrovascular occlusion caused cortical blindness in 7 of 12 patients; carbon monoxide poisoning, trauma, Hodgkin's disease, and progressive subcortical leukoencephalopathy were also reported as causes.

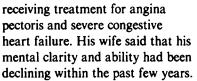
#### **Recent case studies**

It is important to note that the foregoing studies were completed prior to the 1960s-before effective methods of cardiopulmonary resuscitation were developed. More recent reports<sup>2.6-8</sup> of cortical blindness in adults and the three following cases-which occurred in a two-year period at two community hospitals-may indicate an incidence higher than originally suspected and a shift in the primary etiology from cerebrovascular accidents to severe hypoxic or hypotensive episodes occurring during cardiorespiratory arrest.

CASE 1—A 71-year-old man was brought to the emergency department of Borgess Hospital, Kalamazoo, Michigan, in October 1975 by ambulance for evaluation of increasing dyspnea over the previous 24 hours. The patient had been *continued*  Cortical blindness should be suspected when a patient with normal pupillary reflexes complains of blindness even though the results of an ophthal-moscopic examination are normal.

#### Walter G. Drymalski

Dr Drymalski is a fellow in rheumatology, University of Michigan Medical School, Ann Arbor, and Wayne County General Hospital, Eloise, Michigan. He was a resident in internal medicine, Bronson Methodist and Borgess hospitals, Southwestern Michigan Area Health Education Center, Kalamazoo. His current research interests are in hypertrophic pulmonary osteoarthropathy.



On physical examination, the patient did not respond to commands and had peripheral cyanosis. Pupils were dilated but reactive; systolic blood pressure was 50 mm Hg with supraventricular tachycardia in a rate of 130 to 150/min. Cardiopulmonary resuscitation was begun.

The patient improved steadily, and in 72 hours the resuscitation was stopped. At that time he was awake although disoriented to place and time. He also complained of blindness. Initially, the complaint was attributed to the patient's dementia; therefore, complete assessment was delayed for several days.

Over the next few days, the patient began to deny his blindness, although examination revealed



only gross perception of light and complete inability to identify objects or colors. A neurologic consultant found normal optic fundi and cranial nerves, bilateral carotid bruits on auscultation, and no nystagmus in response to optokinetic stimulation with a rotating drum. He also found the patient to be disoriented to place and time and noted occasional visual hallucinations.

In other respects, the patient improved, and he was discharged on a regimen for treatment of congestive heart disease. Within a month he was readmitted for exacerbation of congestive heart failure. No change in blindness was noted at that time, and although he was oriented and cooperative, visual field examination was not requested.

CASE 2—A 41-year-old man was admitted to Bronson Methodist Hospital, Kalamazoo, Michigan, in July 1976 for essential hypertension. He had adult-onset type 1 (insulin-dependent) diabetes mellitus and was taking 40 units of isophane (NPH) insulin daily. He had had a myocardial infarction two years before this admission and a right hemispheric cerebrovascular accident one year before.

Blood pressure with the patient seated was 180/120 mm Hg, carotid pulses were full and without bruits, and heart rate was regular with an atrial gallop. Neurologic examination showed increased deep tendon reflexes on the left side and slight ataxia. Methyldopa, propranolol, and furosemide were administered.

On the third day, the patient became agitated and complained of being unable to see. Examination at that time showed reactive pupils, normal fundi, blood pressure of 140/100 mm Hg, and blood glucose level of 105 mg/dl. The complaint of blindness was not believed initially, and chlorpromazine was administered to control agitation.

By the fourth day, the patient was disoriented and more agitated. Left-sided hemiparesis developed; blood pressure was 110/60 mm Hg. The patient stated that he could see, although when examined he was unable to identify objects or visually follow their movement.

The neurologic consultant agreed that cortical blindness, as manifested by intact pupillary reflexes and absence of response to optokinetic stimulation, may have Other than complaints of blindness, the most common symptom of cortical blindness is denial of blindness, known as Anton's syndrome.

occurred. A radioisotope brain scan showed decreased uptake in the left parietal lobe and several small foci of decreased uptake in the right posterior parietal lobe.

After prolonged hospitalization for intensive physical therapy, the patient recovered only minimal use of his left leg and no apparent use of his left arm, although evaluation was complicated by his denial of that arm. The patient continued to deny his blindness but was unable to consistently identify colors or figures. Visual field examination was not done during his hospitalization.

During more than one year of follow-up, no significant change in the patient's blindness was noted.

CASE 3—A 65-year-old man was brought to the emergency department at Borgess Hospital in September 1977 with a complaint of rapid onset of dyspnea while resting. Two days previously, digoxin therapy had been started for probable aortic stenosis with dyspnea on exertion. At that time, the patient also had had symptoms consistent with angina pectoris, and an ECG had shown left bundlebranch block.

Shortly after arrival, the patient had respiratory arrest, and intubation and resuscitation were begun. When his condition stabilized, he was transferred to the intensive care unit. While being examined there, he opened his eyes in response to noises or voices but would not follow commands. Within three hours irregular, unstereotyped, jerking movements of the extremities developed. The neurologic consultant, who felt these signs were consistent with myoclonic-type seizures, advised against immediate treatment because the movements were infrequent. Cranial nerves were thought to be intact, and no focal abnormalities were identified.

The next day, the patient appeared more alert. He followed simple commands but with a tendency toward perseveration. He also began talking at intervals and, during one of these periods, complained of blindness.

On examination, the pupils reacted to light, but there was no blinking response to threatening gestures. Optokinetic stimulation also produced no response.

The following day, he no longer complained of blindness and, at one time, seemed successful in identifying objects. However, later the same day, an EEG showed diffuse slowing, particularly in the occipital regions. Photic stimulation produced no response on the EEG.

Five days after admission, the patient still did not respond to optokinetic stimulation, and the EEG, which showed some increase in background slow-wave activity, still indicated no significant change with photic stimulation.

The ophthalmologic consultant found a cholesterol embolus in the

right temporal field but believed that the fundi were otherwise normal. Visual field examination was not performed because the patient was unable to cooperate.

Eight days after the patient's admission, gastrointestinal bleeding developed, followed by renal failure. Peritoneal dialysis was unsuccessful, and the patient died six days later.

Autopsy showed aortic stenosis with marked left ventricular hypertrophy, acute bronchopneumonia, and acute ischemic colitis. Grossly, the brain was normal, but microscopic study revealed focal laminar vacuolization and neuronal degeneration throughout the cortex, including sections taken from calcarine sulci.

#### **Clinical presentation**

Cortical blindness should be suspected when a patient with normal pupillary reflexes complains of blindness even though the results of an ophthalmoscopic examination are normal. Too often the complaint of blindness is attributed to dementia or hysteria, which can delay diagnosis, as in cases 1 and 2.

Cortical blindness is far more difficult to diagnose in patients who cannot easily communicate their loss of sight, such as the very young, the very debilitated, and those on respirators. In addition, diagnosis is difficult in patients with Anton's syndrome—a form of anosognosia. In these types of pacontinued Besides the ophthalmoscopic examination, other essential diagnostic tests include bedside examinations, such as optokinetic stimulation, EEG with photic stimulation, and visual field assessment.

#### Table 1. Characteristics of cortical blindness

Classic triad Blindness, evidenced by Failure to react to threatening gestures No response to optokinetic stimulation Consistent EEG findings Intact pupillary reflexes Normal fundi

Additional findings Unformed visual hallucinations Denial of blindness (Anton's syndrome)

tients, close and consistent clinical observation will detect decreased visual attentiveness and sightless behavior.

In about 50% of patients whose blindness is caused by emboli or other occlusive vascular events, history of significant prodromes is evident. The most frequent complaint is transient visual impairment; less often the complaint is vertigo, and least often brief loss of consciousness suggesting vertebrobasilar insufficiency.<sup>4</sup> In addition, while loss of vision is usually acute, it may also occur progressively over several days in a minority of patients.<sup>4</sup>

Of the clinical symptoms associated with cortical blindness, none is constant or pathognomonic. One of the most common is Anton's syndrome. This syndrome, which in the past was considered to occur only with cortical blindness, has also occurred with blindness caused by damage to the retina or optic nerve.<sup>9</sup> As indicated in cases 1 and 2, in which denial of blindness occurred only after an initial complaint of blindness, Anton's syndrome may, at times, arise from the patient's confusion at the hospital staff's failure to confirm his or her initial complaint.

Spatial disorientation, loss of visual imagery, loss of color vision, and visual agnosia also have been associated with cortical blindness, although they were not common occurrences in the Symonds and Mackenzie study,<sup>4</sup> and their incidence in cases occurring after cardiorespiratory arrest has not been reported. In addition, these phenomena were absent in two of the three cases cited here.

Finally, visual hallucinations may accompany cortical blindness. As cases 1 and 2 demonstrate, it may be difficult to distinguish these hallucinations from those of preexistent or concurrent dementia. In case 1 the patient had probable preexistent dementia, which could have been exacerbated by his hospitalization and loss of sight, causing more obvious confusion and hallucination. In case 2 onset of visual hallucinations and cortical blindness occurred simultaneously with onset of abnormal behavior.

#### **Physical examination**

Specific bedside examinations are necessary to confirm cortical blindness in patients with normal pupillary responses to light and normal ophthalmoscopic findings (ie, no sign of optic atrophy, generalized retinal degeneration, or papilledema). Table 1 lists classic characteristics of the disorder. One of the most common is the absence of blinking as a reflex to threatening gestures made by the examiner. Also common is the absence of blinking when a bright light is shone on the face of a patient who has been in semidarkness. The latter response may occasionally be normal in the absence of significant retained cortical vision, but also may be the first response to return during recovery.

Optokinetic stimulation tests for nystagmus should be given to conscious patients. These tests involve use of a revolving drum with alternating dark and light vertical stripes or pulling a long piece of material with alternating designs across the patient's visual field. In patients with normal pupillary reflexes, cortical blindness is confirmed if one of these maneuvers does not produce nystagmus. The hysterical patient cannot voluntarily suppress nystagmus, while the patient whose blindness is caused by other factors will not have normal pupillary reflexes.

EEG with photic stimulation has been proposed as a diagnostic and prognostic test for cortical blindness.<sup>2,10</sup> Responses consistent with cortical blindness are failure to suppress background activity with photic stimulation and the absence *continued*  After cardiorespiratory arrest, the most common visual field defect is loss of central vision with varying degree of peripheral sparing.

of short-latency visually evoked response waves.<sup>2.3.5</sup> However, studies have not shown any consistent ability for this procedure to predict recovery of sight.<sup>3</sup>

In the cooperative patient, assessment of visual fields may be one of the most useful methods for follow-up of cortical blindness. Patterns of visual field loss may also delineate the pathogenesis.

In their study, Symonds and Mackenzie<sup>4</sup> found that the most common visual field pattern (in 27 of 58 cases) was recovery of central vision with continued absence of any peripheral vision. Only four patients demonstrated loss of central vision with partial or complete sparing of peripheral vision. The others either remained totally blind (14) or regained varying amounts of central and peripheral vision (13). The authors noted that the area of central vision in the striate cortex is closest to the occipital poles. They proposed that since the occipital poles receive a rich collateral supply from the "watershed" of the middle and posterior cerebral arteries, they most likely would be spared from specific occlusive events occurring within the occipital regions.

It is important to note that none of the cases discussed by Symonds and Mackenzie included histories of cardiac or respiratory arrest with subsequent resuscitation. Other studies have shown that the cerebrovascular "watershed" areas

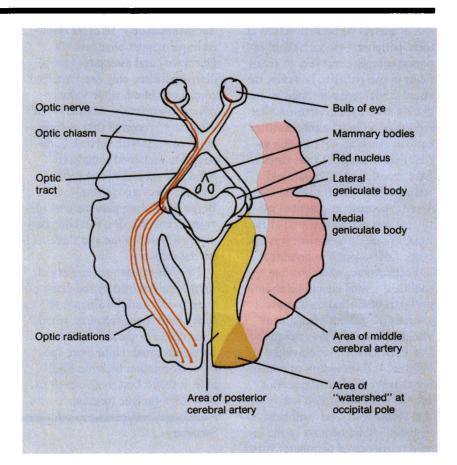


Figure 1. "Watershed" area of middle and posterior cerebral arteries shown in view of optic pathways exposed from below.

are usually the first to be affected by generalized hypoxia because they are the farthest from the origins of cerebral circulation<sup>11</sup> (figure 1). The conclusion seems obvious: In cortical blindness occurring after cardiorespiratory arrest, the most common visual field defect is loss of central vision with varying degrees of peripheral sparing, rather than the preservation of central vision seen with focal occlusive events in the posterior or middle cerebral arteries.

Unfortunately, there are not an adequate number of case reviews in recent literature to document this hypothesis. Hoyt and Walsh<sup>6</sup> reported a case of cardiac arrest in which visual field testing showed *continued*  The majority of patients with cortical blindness have at least some recovery of sight, but the extent of recovery appears to be determined by the cause of the disorder. The amount of time for recovery has not been determined.

loss of central vision with return of some peripheral vision. Sabah<sup>8</sup> reported complete recovery of visual fields in two patients; however, the visual fields were not tested immediately after cardiac arrest. Other reports of occipital blindness after cardiac arrest do not mention visual field findings.<sup>7</sup> In the cases I have reported here, visual field testing was not performed in cases 1 and 2 because its potential usefulness was not appreciated at the time; in case 3 the patient was not cooperative enough to be tested.

Furthermore, when possible, visual field testing should be an integral part of evaluation because it assesses the patient's functional visual status and allows rehabilitation to be tailored to individual abilities. It is important to remember that the patient who retains central vision may have nearly normal visual acuity, but will behave as though blind because of the severe restriction of peripheral visual fields. Such a patient may act totally blind and be treated as such, yet potential for rehabilitation is far greater than that of the patient with no sight preserved.

#### Prognosis

Prognosis for patients with cortical blindness is hopeful. Two thirds or more of those affected will have at least some recovery of sight, although this may not be appreciated without follow-up visual field examination. Only 14 of Symonds and Mackenzie's<sup>4</sup> 58 cases resulted in permanent blindness. Of Duchowny and associates'<sup>3</sup> six pediatric patients, only one remained completely blind, while three recovered completely. Abraham and associates<sup>10</sup> reported that of three adult patients with cortical blindness, one remained completely blind, one recovered completely, and one recovered partially. Of Bergman's<sup>5</sup> seven patients who survived acute episodes, two had complete return of vision and three had partial recovery.

To what extent the recovery of sight is determined by the cause of the disorder and the amount of time required for recovery, whether complete or partial, have not been delineated. Until more specific information becomes available, it seems best to give each patient an optimistic forecast.

#### Summary

Once thought to be rare, cortical blindness today appears to occur more frequently. (Three cases described here occurred within a twoyear period.) In addition, the disorder may have undergone a shift in etiology since the development of effective means of cardiopulmonary resuscitation within the past two decades. Severe hypoxic or hypotensive episodes occurring during cardiorespiratory arrest may have become a more common cause of cortical blindness in adults than cerebrovascular accidents. Prognosis for recovery of at least partial sight is good, although proper visual testing is required to determine the extent of improvement. FCM

Address reprint requests to Walter G. Drymalski, MD, Department of Internal Medicine, Wayne County General Hospital, Eloise, MI 48185.

#### References

Griffith JH, Dodge PR. Transient blindness following head injury in children.
 N Engl J Med 1961;278:648-51
 Barnet AB, Manson JI, Wilmer E. Acute cerebral blindness in children: six cases studied clinically and electrophysiologically.

studied clinically and electrophysiologically. Neurology 1970;20:1147-56 3. Duchowny MS, Weiss IP, Majlessi H, et

3. Duchowny MS, wess IP, Majiessi H, et al. Visual evoked responses in childhood cortical blindness after head trauma and meningitis: a longitudinal study of six cases. Neurology 1974;24:933-40

4. Symonds C, Mackenzie I. Bilateral loss of vision from cerebral infarction. Brain 1957; 80:415-54

5. Bergman PS. Cerebral blindness. Arch Neurol 1957;78:568-84

6. Hoyt WF, Walsh FB. Cortical blindness with partial recovery following acute cerebral anoxia from cardiac arrest. Arch Ophthalmol 1958;60:1061-9

7. Weinberger HA, vanderWoude R, Maier HC. Prognosis of cortical blindness following cardiac arrest in children. JAMA 1962;179:126-9

8. Sabah AH. Blindness after cardiac arrest. Postgrad Med J 1968;44:513-6
9. Redlich FC, Dorsey JF. Denial of blindness by patients with cerebral disease. Arch Neurol 1945;53:407-17

10. Abraham FA, Melamed E, Levy S. Prognostic value of visual evoked potentials in occipital blindness following basilar artery occlusion. Appl Neurophysiol 1975;38: 126-35

11. Brierly JB. Cerebral hypoxia. In: Blackwood W, Corsellis JA, eds. Greenfield's neuropathology. 3rd ed. London: E Arnold, 1976:56-79