

nously, then 6 mg every 6 hours) have been thought to aid in the survival of patients with AHL.^{3,4} More recently, plasma exchange was used successfully in one patient with neurologic sequelae of mycoplasmal pneumonia.¹⁷ Until now, to our knowledge the use cyclophosphamide has not been reported in the treatment of AHL. Its effect in this patient remains uncertain. Effective treatment probably consists of a combination of early diagnosis based on clinical suspicion, neuroimaging, brain biopsy, life support with reduction of elevated ICP in the critical stage, and early, sustained treatment with appropriate immunosuppressants.

References

1. Hurst EW. Acute hemorrhagic leukoencephalitis: a previously undefined entity. *Med J Aust.* 1941;2:1-6.
2. Beham PO, Moore MJ, Lamarche JB. Acute necrotizing hemorrhagic encephalopathy. *Postgrad Med.* 1973;54:154-160.
3. Byers RK. Acute hemorrhagic leukoencephalitis: report of three cases and review of the literature. *Pediatrics.* 1975;56:727-735.
4. Foley JM, Kane CA. Clinical pathologic conference. *Neurology.* 1957;7:135-141.
5. Cox WS, Luse SA. Acute hemorrhagic leukoencephalitis: a clinical and electron-microscopic report of two patients treated with surgical decompression. *J Neurosurg.* 1963;20:584-596.
6. Gosztanyi G. Acute hemorrhagic leukoencephalitis (Hurst's disease). In: Vinken PJ, Brun GW, eds. *Handbook of Clinical Neurology.* Amsterdam, the Netherlands: Elsevier Science Publishers; 1978;3(pt 2):587-604.
7. Lander H. Acute hemorrhagic leukoencephalitis. *Australas Ann Med.* 1958;7:55-68.
8. Kulich SA. Acute hemorrhagic leukoencephalitis: report of a case and review of the literature. *Boston Med Q.* 1960;11:120-130.
9. Martins AN, Kempe LG, Harp GJ. Acute hemorrhagic leukoencephalitis with a concurrent primary herpes simplex infection. *J Neurol Neurosurg Psychiatry.* 1964;27:493-501.
10. Yesnick L. Central nervous system complications of primary atypical pneumonia. *Arch Intern Med.* 1956;97:93-98.
11. Fisher RS, Clark AW, Wolinsky JS, Parhad IM, Moses H, Mardiney MR. Postinfectious leukoencephalitis complicating *Mycoplasma pneumoniae* infection. *Arch Neurol.* 1983;40:109-113.
12. Rothstein TL, Shaw CM. Computerized tomography as a diagnostic aid in acute hemorrhagic leukoencephalitis. *Ann Neurol.* 1983;13:331-333.
13. Watson RT, Ballinger WE, Quisling RG. Acute hemorrhagic leukoencephalitis: diagnosis by computed tomography. *Ann Neurol.* 1984;15:611-612.
14. Levine S, Wenk EJ. A hyperacute form of allergic encephalomyelitis. *Am J Pathol.* 1965;47:61-88.
15. Lamarche JB, Beham PO, Segarra JM, Feldman RG. Recurrent acute necrotizing hemorrhagic encephalopathy. *Acta Neuropathol(Berl).* 1972;22:79-87.
16. Kristianson K, Harkmark W, Cohen MM. Acute hemorrhagic encephalitis. *Neurology (Minneapolis).* 1956;6:503-509.
17. Cotter FE, Bainbridge D, Newland AC. Neurological deficit associated with *Mycoplasma pneumoniae* reversed by plasma exchange. *BMJ.* 1983;286:22.

Isolated Congenital Hemianopia Caused by Prenatal Injury to the Optic Radiation

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• Isolated congenital hemianopias are typically caused by developmental abnormalities of the occipital lobe cortex. We describe two patients with an incidental partial hemianopia associated with unilateral periventricular leukomalacia that was acquired prenatally. Magnetic resonance imaging scans suggest a late second or early third trimester unilateral cerebral ischemic event. (*Arch Neurol.* 1991;48:1088-1091)

Isolated congenital occipital lesions with accompanying hemianopia usually involve gray and white matter. Examples include porencephaly¹⁻³; vascular malformations, either isolated or in association with Sturge-Weber-type malformation without cutaneous signs⁴; colpocephaly⁵; and polymicrogyria of the striate cortex.^{6,7} This report documents two cases of congenital hemianopia caused by prenatal injury to the periventricular white matter.

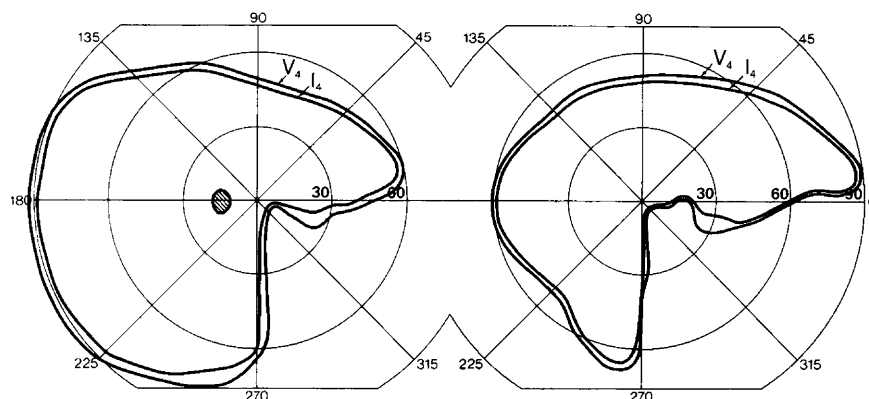
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REPORT OF CASES

CASE 1.—A healthy 21-year-old white woman was found to have an inferior quadrantic field defect during a routine eye examination. She had a history of herpetic keratitis in her right eye. There was no history of prenatal or postnatal illness or neurologic disorder. Her visual acuities were 20/50 in the right eye and 20/20 in the left eye. There were no abnormalities of color vision. Her pupil reflexes were equal bilaterally. Both Goldmann and computerized visual field testing showed a right inferior homonymous quadrantanopia with minor incongruity along the horizontal meridian (Fig 1). There were signs in both retinas of nerve fiber loss corresponding to that seen in homonymous hemiopic atrophy.⁸ Both discs were small in diameter. The right disc showed band atrophy, with loss of nerve fiber layer especially prominent at the nasal disc edge, where the pigment epithelial edge could be seen. The superior and inferior arcuate bundles were well preserved. On the left disc, a full complement of nasal nerve fiber layer could be seen, but temporal pallor and relative loss of the superior and inferior arcuate bundles

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Fig 1.—Patient 1. Right inferior homonymous quadrantic defect demonstrated by Goldmann perimetry.



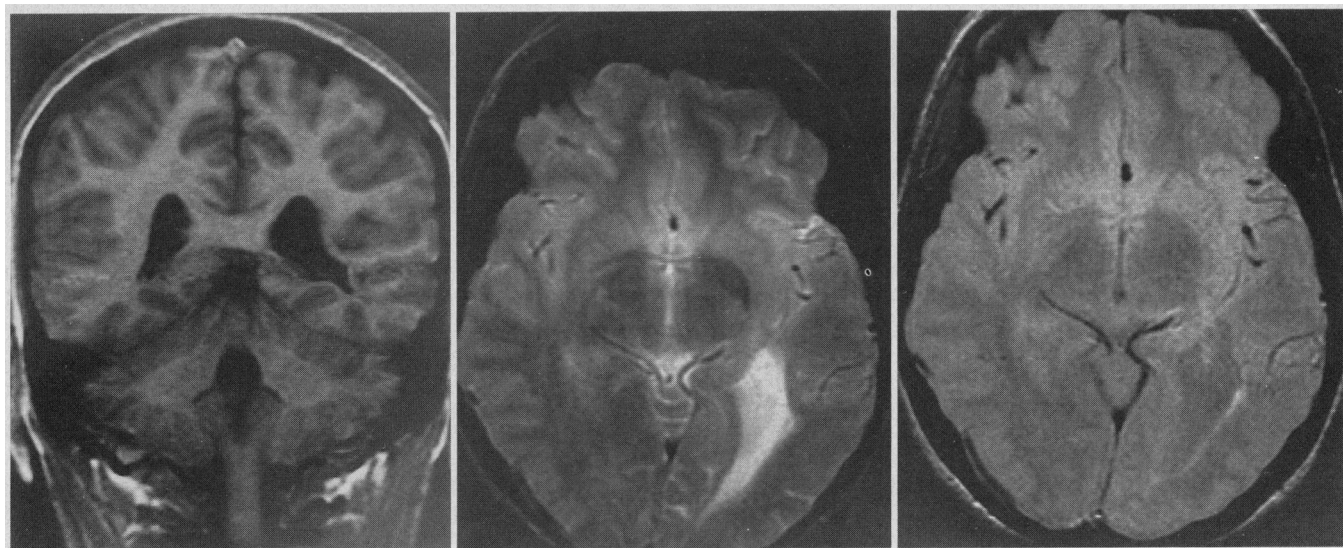


Fig 2.—Patient 1. Left, Coronal T_1 -weighted image at the level of the ventricular trigone shows a nearly complete absence of periventricular white matter inferolaterally on the left. The depths of the cortical gyri nearly about the ventricular surface. Center, Heavily T_2 -weighted axial image shows the expanded trigone and paucity of periventricular white matter. The lateral margin of the ventricle is poorly defined. Right, Spin-density image at the same level as center shows high signal intensity in the periventricular white matter, representing damaged tissue. The presence of this area of high intensity indicates that the damage occurred later than that in patient 2 (compare with Fig 4, right).

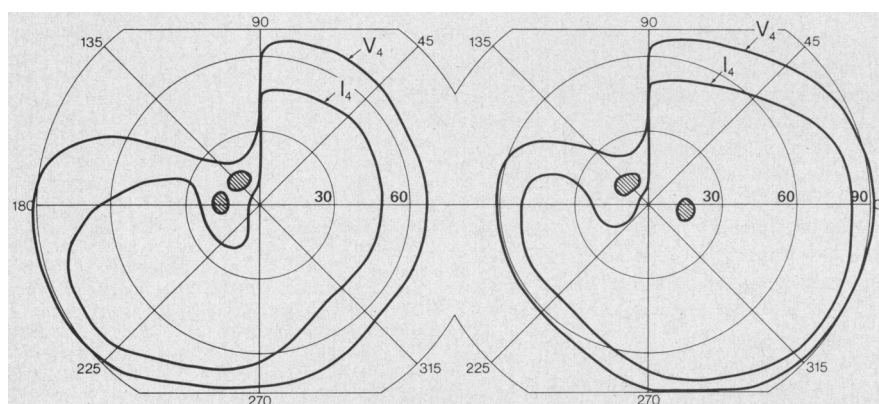


Fig 3.—Patient 2. Left superior homonymous quadrantic defect demonstrated by Goldmann perimetry.

were present.

Magnetic resonance images of the brain (Fig 2) showed ex vacuo enlargement of the trigone and occipital horn of the left lateral ventricle. The close approximation of the cortical gyri, particularly the posterior insular cortex, to the ventricular surface was characteristic of the pattern seen as a result of loss of the intervening white matter. In this location, the white matter is partially composed of the optic radiations. T_2 -weighted images showed a thin strip of high signal intensity in the immediate periventricular white matter indicative of replacement of normal tissue by gliotic white matter.

CASE 2.—A healthy 10-year-old black boy, who was being followed up in the eye clinic for a left esotropia and amblyopia, was found to have a left homonymous hemianopia. There was no significant obstetric or perinatal history. His visual acuities were 20/25 in the right eye and 20/200 in the left eye. The

pupils were 3 mm in diameter, equal, and fully reactive. There was a mild left relative afferent pupillary defect. The patient had a left superior homonymous quadrantanopia (Fig 3). The retinal nerve fiber layer demonstrated a pattern of nerve fiber loss indicating homonymous hemioptic atrophy. The remarkable feature of both discs was that they were large in diameter, with pronounced physiological cups and a nasal entrance of the vessels. The left disc was vertically oval and kidney shaped with band atrophy. There was an atrophic nerve fiber layer at the nasal border of the left disc compared with a full nasal nerve fiber layer adjacent to the right disc.

Computed tomography of the head showed enlargement of the right lateral ventricle, most notably the trigone, as a result of loss of periventricular white matter. Magnetic resonance imaging with gadolinium-pentetic acid (Fig 4) showed the same enlargement of

the ventricle and more clearly showed the close approximation of the depths of the cortical gyri to the ventricular wall. T_2 -weighted images showed normal intensity of the periventricular white matter. No enhancement was seen after administration of intravenous contrast.

COMMENT

We have described two cases of unilateral white matter injury in the region of the optic radiations causing isolated partial hemianopias. In both cases the ophthalmologic evidence of transsynaptic nerve fiber degeneration establishes the occipital injury as a prenatal event.²² The occurrence of such noticeable retinal nerve fiber layer and optic disc changes is somewhat unexpected: the predicted loss of nerve fibers by transsynaptic degeneration from a lesion that causes a quadrantanopia would be about 25% from each optic nerve. We have no explanation for the fact that both patients' homonymous field defects were quadrantic, while the magnetic resonance imaging scans showed diffuse, not focal, thinning of the visual radiation in the involved occipital lobes.

The affected regions of both brains have magnetic resonance imaging characteristics that fit those reported for end-stage periventricular leukomalacia.^{10,11} These are (1) ventriculomegaly, sometimes with irregular outline of the ventricular surface; (2) reduced amounts of periventricular white matter at the trigone and, in more severe cases, the entire centrum semiovale; (3)

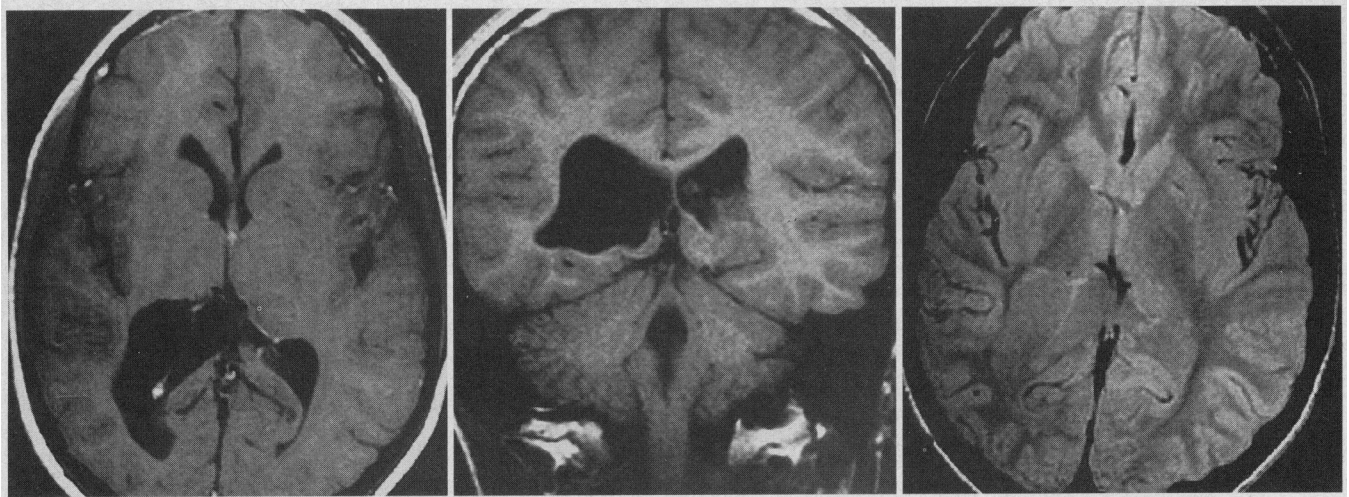


Fig 4.—Patient 2. Left, Axial T₁-weighted image after infusion of intravenous contrast shows enlarged frontal horn and trigone of the right lateral ventricle. Note the paucity of white matter between the cortical gyri and the ventricular surface on the right side compared with the left. No contrast enhancement is apparent. Center, Coronal T₁-weighted image at the level of the ventricular trigones shows that the absence of periventricular white matter is more marked inferiorly at the level of the optic radiations. Right, Axial spin-density image at the level of the ventricular trigones shows an absence of high signal intensity in the periventricular white matter (compare with Fig 2, right). This implies an earlier gestational event.

deep and prominent cortical sulci with subcortical white matter approaching the ventricles with little interposed white matter; and (4) prolonged T₂ relaxation time in the immediate peritrigonal white matter, sometimes extending anteriorly along the ventricular body.

Periventricular leukomalacia occurs most commonly in premature infants who also have other complications including hyaline membrane disease, persistent fetal circulation, and germinal matrix hemorrhage.^{12,13} Both pathological and neuroimaging studies have shown a characteristic progression that begins with edema, followed by coagulation necrosis and eventually cavitation, usually in 3 to 6 weeks.¹²⁻¹⁴ The cysts usually collapse, the affected brain exhibiting gliosis and reduction in the quantity of white matter in the periventricular region. The ependymal separation between the cavities and the ventricle may break down, with consequent incorporation of the cavities into the ventricle.

Since periventricular leukomalacia occurs most commonly in the peritrigonal region,¹⁵ through which the geniculocalcarine radiations course, it is not surprising that patients with this condition commonly have visual impairment.^{16,17} However, this is usually seen in the context of a premature infant with bilateral defects. Several authors have described unilateral periventricular leukomalacia¹⁸⁻²¹; however, all have concentrated on motor dysfunction and none have described visual impairment.

The pathogenesis of most cases of periventricular leukomalacia is now widely accepted as being the result of ischemia in the immature brain.^{11,22,23} The “watershed areas” of the premature brain are located in the periventricular region.²⁴ At about 36 weeks’ gestational age, the watershed areas begin to move peripherally, eventually reaching the subcortical white matter and cerebral cortex at term. It is a logical extension of this concept to postulate that unilateral periventricular leukomalacia is most likely to be the result of in utero hypoperfusion of a single hemisphere.

Gilles et al²⁵ have shown that the glial response to brain injury is minimal prior to about 26 weeks’ gestational age. Barkovich and Truwit¹ have further shown that asphyxial brain injury prior to 28 weeks’ gestational age shows minimal periventricular T₂ prolongation. The area of prolonged T₂ relaxation time adjacent to the enlarged trigone in patient 2 indicates an injury between about 28 and 34 weeks’ gestational age, whereas the absence of periventricular prolonged T₂ relaxation in patient 1 indicates an earlier injury, most likely during the second half of the second trimester.

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References

1. Remillard GM, Ethier R, Andermann F. Temporal lobe epilepsy and perinatal occlusion of the posterior cerebral artery: a syndrome analo-

gous to infantile hemiplegia and a demonstrable etiology in some patients with temporal lobe epilepsy. *Neurology*. 1974;24:1001-1009.

2. Hoyt WF. Congenital occipital hemianopia. *Neuro-ophthalmol Jpn*. 1985;2:252-259.

3. Novakovic P, Taylor DSI, Hoyt WF. Localising patterns of optic nerve hypoplasia—retina to occipital lobe. *Br J Ophthalmol*. 1988;72:176-182.

4. Andriola M, Stolfi J. Sturge-Weber syndrome: report of an atypical case. *AJDC*. 1972;123:507-510.

5. Garg BP. Colpocephaly: an error of morphogenesis? *Arch Neurol*. 1982;39:243-246.

6. Tyehsen L, Hoyt WF. Relative afferent pupillary defect in congenital occipital hemianopia. *Am J Ophthalmol*. 1985;100:345-346.

7. Taylor DC, Falconer MA, Bruton CJ, Corsellis JAN. Focal dysplasia of the cerebral cortex in epilepsy. *J Neurol Neurosurg Psychiatry*. 1971;34:369-387.

8. Hoyt WF, Rios-Montenegro EN, Behrens MM, Eckelhoff RJ. Homonymous hemioptic hypoplasia: fundoscopic features in standard and red-free illumination in three patients with congenital hemiplegia. *Br J Ophthalmol*. 1972;56:537-545.

9. Miller NR, Newman SA. Transsynaptic degeneration. *Arch Ophthalmol*. 1981;99:1654.

10. Flodmark O, Lupton B, Li D, et al. MR imaging of periventricular leukomalacia in childhood. *AJNR*. 1989;10:111-118.

11. Barkovich AJ, Truwit CL. Brain damage from perinatal asphyxia: correlation of MR findings with gestational age. *AJNR*. 1990;11:1087-1096.

12. Volpe JJ. Hypoxic-ischemic encephalopathy: neuropathology and pathogenesis. In: *Neurology of the Newborn*. 2nd ed. Philadelphia, Pa: WB Saunders Co; 1987:209-235.

13. Friede RL. *Developmental Neuropathology*. 2nd ed. New York, NY: Springer-Verlag NY Inc; 1989:69-97.

14. De Reuck J, Chattha AS, Richardson EP. Pathogenesis and evolution of periventricular leukomalacia in infancy. *Arch Neurol*. 1972;27:229-236.

15. Banker BQ, Larroche J-C. Periventricular leukomalacia of infancy. *Arch Neurol*. 1962;7:386-410.

16. Lambert SR, Hoyt CS, Jan JE, Barkovich AJ, Flodmark O. Visual recovery from hypoxic cortical blindness during childhood: computed to-

mography and magnetic resonance imaging predictors. *Arch Ophthalmol*. 1987;105:1371-1377.

17. Flodmark O, Jan JE, Wong PKH. CT of the brains of children with cortical visual impairment. *Dev Med Child Neurol*. 1990;32:611-620.

18. Wiklund L-M, Uvebrant P, Flodmark O. Morphology of cerebral lesions in children with congenital hemiplegia: a study with computed tomography. *Neuroradiology*. 1990;32:179-186.

19. Molteni B, Oleari G, Fedrizzi E, Bracchi M. Relation between CT patterns, clinical findings and etiological factors in children born at term, affected

by congenital hemiparesis. *Neuropediatrics*. 1987;18:75-80.

20. Claeys V, Deonna T, Chrzanowski R. Congenital hemiparesis: the spectrum of lesions. *Helv Paediatr Acta*. 1983;38:393-455.

21. Kotlarek F, Rodewig R, Brull D. CT findings in congenital hemiparesis in childhood and their relation to etiology and prognosis. *Neuropediatrics*. 1981;12:101-109.

22. Pape KE, Wigglesworth JS. *Haemorrhage, Ischaemia, and the Perinatal Brain*. Philadelphia, Pa: JB Lippincott; 1979:69/70. Clinics in Develop-

mental Medicine.

23. Wiklund L-M, Uvebrant P, Flodmark O. CT as an adjunct in etiological analysis of hemiplegic cerebral palsy. *Neuropediatrics*. In press.

24. Takashima S, Tanaka K. Development of cerebrovascular architecture and its relationship to periventricular leukomalacia. *Arch Neurol*. 1978;35:11-16.

25. Gilles FH, Averill DR Jr, Kerr C. Changes in neonatally induced cerebral lesions with advancing age. *J Neuropathol Exp Neurol*. 1977;36:666-684.

Visual Hallucinations and Racing Thoughts on Eye Closure After Minor Surgery

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• **Two hours after minor surgery under local anesthesia, a 67-year-old man developed vivid visual hallucinations that were present only when he closed his eyes. After lasting 4 hours, the hallucinations were succeeded by racing thoughts, lasting 2 hours, and then by illusionary movements of his body. The reaction was attributed to the effect of lidocaine. The case resembles one previously reported. (Arch Neurol. 1991;48:1091-1092)**

The occurrence of visual hallucinations on eye closure in a fully alert patient is a rarely reported phenomenon. Recently, one case was described in which the hallucinations were attributed to atropine toxicity and lasted 11 days.¹ The present case bears a close resemblance; but, in addition to visual hallucinations, the patient experienced "racing thoughts" (his term) and hallucinations of body movement on eye closure. The symptoms began shortly after a minor operation that was performed under local anesthesia.

Abnormal mental events are of interest, not only as they aid in clinical interpretation, but also as they extend our knowledge of the brain's behavioral capability.

REPORT OF A CASE

A 67-year-old hypertensive physician reported a 15-minute spell of weakness of the right side of the face and the right hand. Results of neurological examination were normal, as was a cranial computed tomographic scan. Angiography showed stenosis of both internal carotid arteries to less than 1-mm residual lumen, worse on the right side. A right-sided carotid endarterectomy was attempted, but patency could not be restored, and the internal carotid artery was

ligated. Immediate recovery was uneventful; but, 24 hours later, on removing the drain from the wound, bleeding occurred into the neck, forming a mass that displaced the trachea. The blood clot was promptly evacuated using 15 mL of 0.01% lidocaine, with 0.005% phenylephrine hydrochloride as the local anesthetic. The only additional medications were codeine, 60 mg, given orally 2 hours before the operation; cefazolin sodium, 1 g; and labetalol, 5 mg, administered twice during the procedure, which was uncomplicated and lasted about 1 hour. Cerebral hypoxia did not occur. The labetalol temporarily replaced atenolol, which the patient had been taking for about 6 years.

At 12:30 PM, two hours after surgery, while drowsing, the patient began to experience visual hallucinations; these continued for about 4 hours. According to his daughter, who sat at his bedside as he described the events to her, he was lucid and appropriate at all times. He was first seen by me a few hours after the aberrant ideation had ceased; at this time, his memory of the happenings was still fresh.

The visual hallucinations consisted of four main topics: books and other printed material, street scenes, snowy landscapes, and animals of many kinds. The images followed one another in quick succession, "nonstop with no rhyme or reason," each one lasting 2 or 3 seconds. When he opened his eyes, the visions disappeared. Although he was wide awake and apparently clear mentally, it was for some reason difficult to keep his eyes open. "It was as if they were being forced shut." He was not frightened or alarmed, but was restless and unduly excited; he attributed this to a feeling of being out of control and helpless. The images were colored and projected into the center of space before him, without lateralization. They were silent, and figures showed no facial movements to suggest that they were speaking. The individual hallucinations were entirely unrelated to one another, with no recognizable narrative connection. It was broad daylight, and the room was brightly lit.

The printed matter consisted of books and pages; in addition, handwritten material was present. The colored street scenes were not

recognized as being in any particular city; many people about 1 m tall were rushing across the street from side to side at great speed, dashing hither and thither, creating a bewildering confusion. The streets were lined with buildings on each side. The people seemed to be further away than the surroundings, creating the appearance of miniaturization. There were no autos. The snow scenes stretched for miles over low hills. As the hospitalization was in the winter time, when there was snow, this may have been a determining factor; but, a snow scene could quickly be followed by one without snow. There were no trees. The animals were deformed and repugnant; for example, there were ugly, one-eyed Picasso-like monsters, or, grotesque, nongeometrical shapes that were everchanging. The animals were stationary, as in a picture. There were no recognizable normal animals, such as dogs, cats, horses, or rodents. The coloring was mostly of water colors and pastels, except for white, which was quite bright, and orange, which was vivid.

As the visual hallucinations subsided, they were followed by a most unpleasant episode of "racing thoughts." The patient was not aware of any visual or auditory images. Unconnected thoughts followed one another in quick succession, without any sense. They passed so swiftly, the patient was unable to recognize them. It was his impression that they were about people, rather than things or places. They probably involved family members, including a grandson. Each thought might consist of three staccato bits, only to be interrupted by an entirely different thought with its own few bits, with the whole series being fragmentary and meaningless. Also, the thinking spree was present only when his eyes were closed; this was immediately dispelled on opening the eyes; but, again, there was difficulty in keeping the eyes open in the absence of sleepiness. The episode of rushing thoughts was stranger and more unpleasant than the visual hallucinations, although the thoughts themselves were not frightening or bizarre. The patient agreed that the thoughts might have been ordinary thinking, speeded up to a dizzying degree. In light of the patient's statement

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