Figure 4. The amount of reduction in the exposed ocular surface area in relation to the amount of reduction in exophthalmos after decompression surgery.

Neurological involvement in thrombotic thrombocytopenic purpura (TTP) is frequent. In one series, magnetic resonance imaging revealed brain lesions in 88% of the patients with TTP. The 2 most common cerebral lesions associated with TTP are edema and infarction. Cerebral edema predominantly affects the white matter, but when it affects gray matter in the territory of the posterior cerebral circulation, it may resemble the radiological findings of reversible posterior leukoencephalopathy syndrome (RPLS). We report a case of reversible cortical blindness caused by RPLS in a patient with TTP exacerbation and pre-eclampsia. To our knowledge, there have been only 10 previously reported cases of RPLS in the setting of TTP. We also discuss the role of diffusion-weighted imaging (DWI) in differentiating reversible from irreversible ischemic lesions.

**Case Report.** A 19-year-old, 28-week pregnant white woman came to the hospital with gross hematuria and decreased urine output. Her medical history was significant for recurrent episodes of TTP. Her pregnancy was complicated by 3 prior admissions for TTP exacerbation. Each time she responded to fresh frozen plasma transfusion. Her medications included iron supplements and prenatal vitamins.

At admission her blood pressure was 175/84 mm Hg, and she was afibrile. The remainder of the physical examination results were unremarkable. Laboratory testing re-
revealed a hematocrit level of 27.7%, platelet count of 13 x 10^3/μL and a creatinine level of 2.0 mg/dL (176.8 μmol/L). A peripheral blood test revealed schistocytes. Results of liver function tests and routine coagulation studies were normal. We diagnosed TTP exacerbation, and the patient was given daily fresh frozen plasma transfusions. Her blood pressure was controlled with intravenous labetalol hydrochloride, ranging from 130/70 mm Hg to 140/80 mm Hg. However, on day 3 of the hospital admission, her blood pressure reached systolic levels of 160 to 170 mm Hg and diastolic levels of 80 to 90 mm Hg despite aggressive treatment with labetalol and the addition of hydralazine hydrochloride. Later the same day, she developed a severe headache, nausea, and vomiting, and ultrasonography revealed no fetal cardiac activity. The following day, she noted a sudden loss of vision in both eyes, which she described as total blackness. Her blood pressure at the time of visual loss was 183/96 mm Hg. Results of a cranial computed tomographic image without contrast were interpreted as normal. The visual acuity was hand motions OU. Both pupils were briskly reactive to direct light with no relative afferent pupillary defect. She did not generate nystagmus in either eye with the optokinetic drum. Anterior segment and dilated fundus examination findings were normal. Neurological examination revealed a lethargic, irritable patient with no focal deficits. A cranial magnetic resonance imaging study showed increased signal intensity of the gray and subcortical white matter on fluid-attenuated inversion recovery and T2-weighted images bilaterally within the posterior cerebral artery territories associated with mild mass effect, reflected as narrowing of the sulci in the involved areas. The DWI demonstrated an isointense signal in the same distribution as the T2-signal abnormalities, except for a small increased signal lesion in the right posterior temporal lobe (Figure 1). The following day, after an induced delivery of a stillborn fetus, the visual acuity improved to 20/200 OU, and the blood pressure normalized. At a follow-up examination 5 weeks later, the visual acuity improved to 20/20 OD and 20/25 OS. Results of automated perimetry were normal in both eyes. Repeated magnetic resonance imaging demonstrated resolution of the previous signal abnormalities in the temporal and occipital lobes (Figure 2).

**Comment.** Our patient’s ocular examination results, radiological findings, and clinical course were consistent with RPLS. Previously referred to as hypertensive encephalopathy, RPLS has been associated with a variety of conditions including pre-eclampsia, eclampsia, an adverse

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**Figure 1.** Magnetic resonance imaging study at the time of visual acuity examination measuring hand motions OU. A, Axial fluid-attenuated inversion recovery image through the level of the temporal and occipital lobes demonstrates bilateral asymmetric areas of increased signal intensity involving the gray and subcortical white matter of the posterior temporal and occipital lobes. B, Diffusion-weighted image (DWI) shows all but 1 lesion in the right posterior temporal lobe isointense to the normal-appearing brain parenchyma. The lesion in the posterior temporal lobe (arrow) shows increased signal intensity, suggestive of restricted diffusion. C, The apparent diffusion coefficient (ADC) map does not demonstrate decreased signal intensity in the area of interest (arrow), as would be expected based on the DWI, but rather increased signal intensity suggestive of vasogenic edema. The contradicting findings on the DWI and ADC map are due to the T2 “shine-through” phenomenon.

**Figure 2.** Follow-up magnetic resonance imaging study with visual acuity measuring 20/20 OD and 20/25 OS. Axial fluid-attenuated inversion recovery (A) and diffusion-weighted (B) images obtained 4 weeks after the initial magnetic resonance imaging study showed complete resolution of the previous signal abnormalities.
effect of chemotherapeutic drugs, acute renal insufficiency, and TTP.1,2

The exact mechanism of RPLS remains unclear. Most patients have elevated systemic blood pressure and develop reversible cerebral edema, with cerebral infarction occurring rarely.1,2 Two main theories have been proposed to explain the pathomechanism of the disease. The first suggests an acute rise in systemic arterial blood pressure inducing autoregulatory cerebral vasospasm, which results in ischemia and subsequent cytotoxic edema. When severe enough, the ischemia may progress to infarction.3 The second theory proposes that the acute hypertension results in dysautoregulation and passive vasodilation of the cerebral vasculature, resulting in hyperperfusion and extravasation of fluid into the interstitium (vasogenic edema). Recent radiological studies using DWI have demonstrated that in most patients with RPLS, the cerebral edema is vasogenic in origin and characterized by increased diffusion of water molecules, in contrast to cytotoxic edema, which shows restricted water diffusion.2,4 Furthermore, the reversibility of the lesions and their preference for the posterior circulation with less vasomotor sympathetic innervation supports a vasogenic pathogenesis.

Before the advent of DWI, it was difficult to differentiate between cytotoxic and vasogenic edema in patients with RPLS because they both produce a bright signal on T2-weighted images. On DWI studies, vasogenic edema produces a dark or isointense signal (high diffusion) compared with cytotoxic edema, which produces a bright signal.2,4 However, a bright signal on DWI studies can sometimes be observed with vasogenic edema because of the prolongation of the T2 signal.2 This phenomenon, known as the T2 “shine-through” effect, occurs because the DWI is influenced by water diffusibility and the intrinsic T2 properties of the tissue.3 The difficulty of interpreting this finding is eliminated by generating an apparent diffusion coefficient map, which reflects only the diffusion characteristics of the lesions and is independent of the T2 shine-through effect.2 On apparent diffusion coefficient maps, lesions with restricted water diffusion (ie, cytotoxic edema) are hypointense, and lesions with increased diffusion, as in our case of vasogenic edema, generate normal to increased signal intensity. The distinction between cytotoxic and vasogenic edema is important because the treatment and prognosis are different. Vasogenic edema is reversible with prompt treatment of the underlying cause. Cytotoxic edema, however, suggests an acute infarction and may warrant invasive diagnostic studies and immediate thrombolytic therapy.

In our patient, the mechanism of the cerebral lesions was probably multifactorial, with TTP and preeclampsia as predisposing factors. Widespread endothelial injury, hypertension, and renal failure most likely contributed to the development of vasogenic edema. Treatment with fresh frozen plasma interrupted platelet thrombi formation, preventing further endothelial damage and resulting in improvement of renal function, blood pressure, and vasogenic cerebral edema.

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