PHACE syndrome in a preterm infant
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In a preterm infant (28 weeks), postnatal cranial ultrasound showed unilateral cerebellar hypoplasia. On day 5, a facial erythematous lesion developed, progressing to a segmental hemangioma during the next 2 weeks (figure 1, A and B). PHACE syndrome (posterior fossa anomalies, most commonly located in the mid brain or hindbrain, such as the Dandy-Walker complex and focal dysplasia and/or hypoplasia of the cerebellum, hemangioma, arterial lesions, cardiac abnormalities or coarctation of the aorta, eye or endocrine abnormalities)\(^1\) was suspected. MRI confirmed cerebellar hypoplasia and intracranial hemangioma (figure 2). Magnetic resonance angiography and echocardiogram were normal. Because of obstruction of the visual axis, low-dose atenolol was started (0.5–1.0 mg/kg/d), and continued for 2 years.

Regression of the hemangioma started within the first week of treatment (figure 1, C and D). MRI at 1.5 years showed complete resolution of intracranial hemangioma. Neurodevelopment and ophthalmologic outcome at 2 years were normal. The child developed bilateral conductive hearing loss.
Study funding
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Disclosures
The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

Appendix

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Reference

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Figure 2 Ultrasound and MRI of the brain in a preterm infant with PHACE syndrome

Postnatal ultrasound (A) and MRI at 31 weeks (B, C) show unilateral cerebellar hypoplasia (blue arrows) and ipsilateral periorbital and cerebellopontine angle hemangioma (white arrows).
PHACE syndrome in a preterm infant
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