

# PHACE syndrome in a preterm infant

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*Neurology*® 2020;95:751-752. doi:10.1212/WNL.0000000000010800

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**Figure 1** Evolution of facial hemangioma in a preterm infant with PHACE syndrome



(A) At day 5, erythematous lesion in left frontotemporal segment; (B) at 2 weeks; (C, D) at 6 months and 2 years.

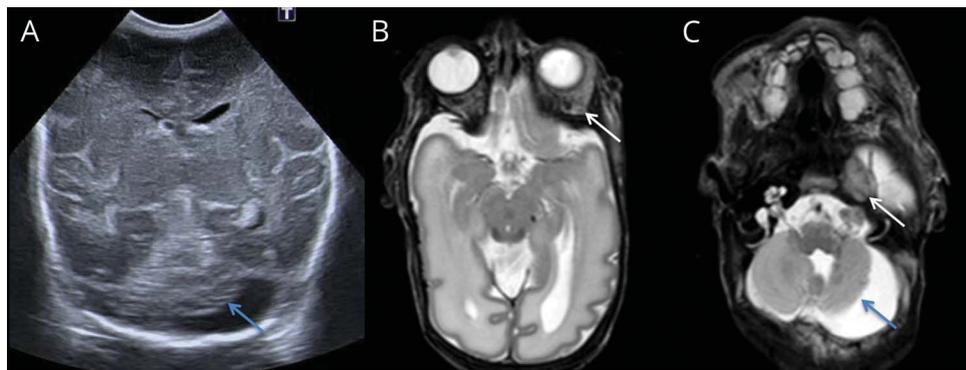
In a preterm infant (28 weeks), postnatal cranial ultrasound showed unilateral cerebellar hypoplasia. On day 5, a facial erythematous lesion developed, progressing to a segmental hemangioma during the next 2 weeks (figure 1, A and B). PHACE syndrome (posterior fossa anomalies, most commonly located in 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)<sup>1</sup> 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.

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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).

### Study funding

No targeted funding reported.

### Disclosures

The authors report no disclosures relevant to the manuscript. Go to [Neurology.org/N](http://Neurology.org/N) for full disclosures.

### Appendix Authors

Name	Location	Contribution
<b>Sylke J. Steggerda, MD, PhD</b>	Department of Paediatrics, Division of Neonatology, Leiden University Medical Centre, the Netherlands	Study concept and design, acquisition of data, neuroimaging, drafted and revised the manuscript

### Appendix (continued)

Name	Location	Contribution
<b>Ratna N.G.B. Tan, MD</b>	Department of Paediatrics, Division of Neonatology, Leiden University Medical Centre, the Netherlands	Study concept and design, acquisition of data, neuroimaging, drafted and revised the manuscript
<b>Peter C.J. de Laat, MD, PhD</b>	Department of Paediatrics, Vascular Anomaly Center, Erasmus Medical Centre, Rotterdam, the Netherlands	Study concept and design, acquisition of data, neuroimaging, drafted and revised the manuscript

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*Neurology* 2020;95:751-752 Published Online before print September 10, 2020

DOI 10.1212/WNL.0000000000010800

**This information is current as of September 10, 2020**

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