PHACE syndrome in a preterm infant

Steggerda, S.J.; Tan, R.N.G.B.; Laat, P.C.J. de

NEUROIMAGES

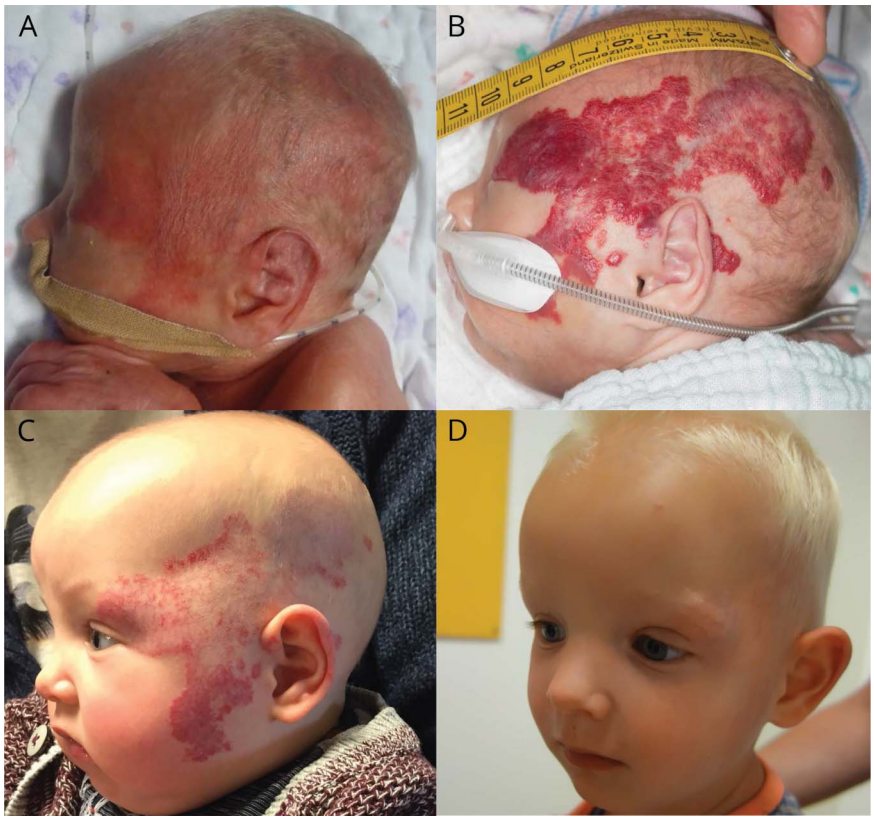
PHACE syndrome in a preterm infant

Sylke J. Steggerda, MD, PhD, Ratna N.G.B. Tan, MD, and Peter C.J. de Laat, MD, PhD

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

Correspondence
Dr. Steggerda
s.j.steggerda@lumc.nl

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.

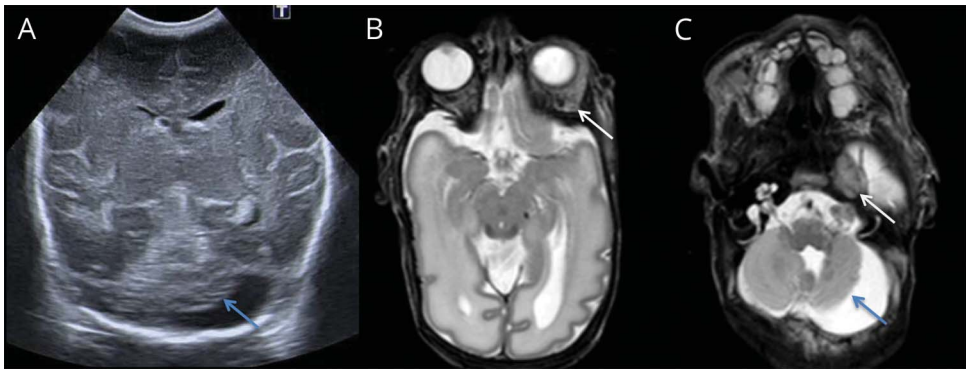
In a preterm infant (28 weeks), postnatal cranial ultrasound showed unilateral cerebellar hypoplasia. On day 5, a facial erythematous lesion developed, progressing to a segmental hemangioma during the next 2 weeks (figure 1, A and B). PHACE syndrome (posterior fossa anomalies, most commonly located in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

From the Department of Paediatrics (S.J.S., R.N.G.B.T.), Division of Neonatology, Leiden University Medical Centre; and Department of Paediatrics (P.C.J.d.L.), Vascular Anomaly Center, Erasmus Medical Centre, Rotterdam, the Netherlands.

Go to Neurology.org/N for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

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 for full disclosures.

Appendix Authors

Name	Location	Contribution
Sylke J. Steggerda, MD, PhD	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
Ratna N.G.B. Tan, MD	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
Peter C.J. de Laat, MD, PhD	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

Reference

1. Garzon MC, Epstein LG, Heyer GL, et al. PHACE syndrome: consensus-derived diagnosis and care recommendations. J Pediatr 2016;178:24–33.

PHACE syndrome in a preterm infant

Sylke J. Steggerda, Ratna N.G.B. Tan and Peter C.J. de Laat

Neurology 2020;95;751-752 Published Online before print September 10, 2020
DOI 10.1212/WNL.0000000000010800