

Pearls & Oysters: Postdural Puncture Headache, Cerebral Sinus Venous Thrombosis, and Reversible Cerebral Vasoconstriction Syndrome in the Peripartum Period

Leonardo Caranzano, MMed, Helene Fenter, MMed, Philippe Maeder, MD, PhD, Patrik Michel, MD, PhD, and Davide Strambo, MD

Neurology® 2022;99:665-668. doi:10.1212/WNL.0000000000201111

Correspondence

Dr. Caranzano
leonardo.caranzano@
chuv.ch

Abstract

We report the case of a 34-year-old female patient complaining of headaches 1 day after childbirth, initially interpreted as postdural puncture headache (PDPH) and treated successfully with an epidural blood patch. Five days later, she presented with an acute proportional right sensorimotor hemisyndrome and a new-onset left-sided headache, attributed to a venous stroke from left-sided cerebral sinus venous thrombosis (CSVT). Simultaneously, we found radiologic signs of reversible cerebral vasoconstriction syndrome (RCVS), considered asymptomatic. We administered anticoagulant therapy to the patient, and she showed full motor recovery at 3-month clinical follow-up. PDPH, CSVT, and RCVS are well-known neurologic complications during the peripartum period. All 3 conditions present with headaches, and headache features may overlap, masking co-occurrence and making accurate diagnosis (differentiation) of these diseases difficult. Each disease can potentially lead to disabling deficits, but all respond to specific treatment. Knowledge of the causes of headaches in the peripartum period, their specific clinical characteristics, and potential complications helps to prioritize and interpret diagnostic tests to offer appropriate therapy.

Pearls

- A large variety of headaches of frequent and rare etiology occur in the peripartum period.
- Excellent history taking and knowledge of clinical headache presentations will guide the choice of diagnostic examinations and their interpretation.
- The simultaneous presence of different headache etiologies in a peripartum patient should be considered.
- Detecting co-occurrence should prompt targeted treatment to reduce chances of complications.

Oysters

- New-onset headache in the peripartum period must be considered a red flag for secondary headaches.
- Proportional hemiparesis of upper and lower limbs cannot be explained by a small arterial cortical stroke as the respective limb representations are vascularized by 2 different arteries, the middle and anterior cerebral arteries.

Case Report

A 34-year-old woman without a previous medical history experienced orthostatic headaches following a vaginal delivery with epidural anesthesia, diagnosed as postdural puncture headache (PDPH). Because the patient was unable to care for her baby because of the pain, following

From the Department of Clinical Neurosciences (L.C., H.F., P. Michel, D.S.), Neurology Service, and Department of Medical Radiology (P. Maeder), Radiodiagnostic and Interventional Radiology Service, Lausanne University Hospital and University of Lausanne, Switzerland.

Go to [Neurology.org/N](https://www.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.

current recommendations for the treatment of PDPH in the obstetrics setting,^{1, 2} early epidural blood patches (EBPs) were performed, which led to headache resolution within 72 hours. No imaging or measurement of CSF opening pressure was performed.

Five days after the delivery, the patient presented to our emergency department because of sudden onset of paresthesia in her right arm and leg followed by weakness in these same extremities. Thirty minutes later, a progressive moderate severity headache appeared that lateralized on the left side of the head.

On physical examination, 180 minutes after symptom onset, the patient had mild paresis of the right arm and leg. Vitals signs were normal, with no indication of elevated blood pressure at any time. Fundoscopic examination findings were normal. Brain MRI showed moderate diffusion restriction and hypoperfusion in the left precentral gyrus, interpreted as an acute arterial ischemic stroke (Figure 1, A and B). MR angiography (MRA) revealed irregularities of multiple segments of the intracranial arteries without significant stenosis or occlusion (Figure 2A). We started IV thrombolysis with rtPA at 225 minutes after symptom onset, with no clinical improvement.

The following day, control MRI detected a parenchymal hemorrhage in the left precentral gyrus within the primary ischemic territory, without mass effect or clinical worsening. The MRI also showed venous thrombosis of a left frontal cortical vein (actually present on the baseline imaging) (Figure 1C) and a new thrombosis of the left transverse sinus. MRA confirmed the persistent irregularities of the intracranial arteries (bilateral middle cerebral artery and left anterior cerebral artery), verified by transcranial Doppler (TCD) examination that found increased flow velocity indicating more than 50% stenosis of the right anterior cerebral artery (Figure 2B).

Our final diagnosis for the acute cortical lesion was a venous infarct, as the lesion was adjacent to the cortical vein thrombosis and the distribution did not respect an arterial territory

but involved both the superficial middle and anterior cerebral artery territories. Further workup included lumbar puncture (LP) and blood examinations for prothrombotic state and vasculitis, and findings of both examinations were normal. Although a second LP carried the risk of aggravating the PDPH, due to the combination of intracranial arterial stenoses, small hemorrhagic infarctions, and cerebral venous thromboses, we considered it vital to exclude possible vasculitis. Indeed, a diagnosis of vasculitis would have required other therapeutic management, with delay or misdiagnosis potentially leading to a negative effect on patient prognosis. The headache did not increase after the second LP.

We diagnosed the arterial irregularities as reversible cerebral vasoconstriction syndrome (RCVS). We administered oral anticoagulation with 5 mg of apixaban twice a day but gave the patient no drugs for vasospasms, considered asymptomatic.

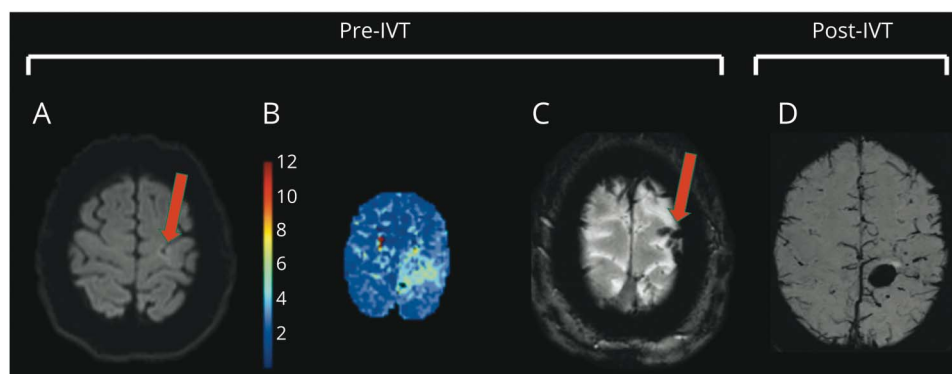
Serial TCD images showed spontaneous regression of the intracranial stenoses, and after 8 days, we discharged the patient home with no neurologic deficits. Repeat MRI 3 months later showed a hemorrhagic scar at the site of the venous infarction, no new lesions, and complete resolution of the cerebral sinus venous thrombosis (CSVT) and arterial abnormalities. Anticoagulation was stopped.

Discussion

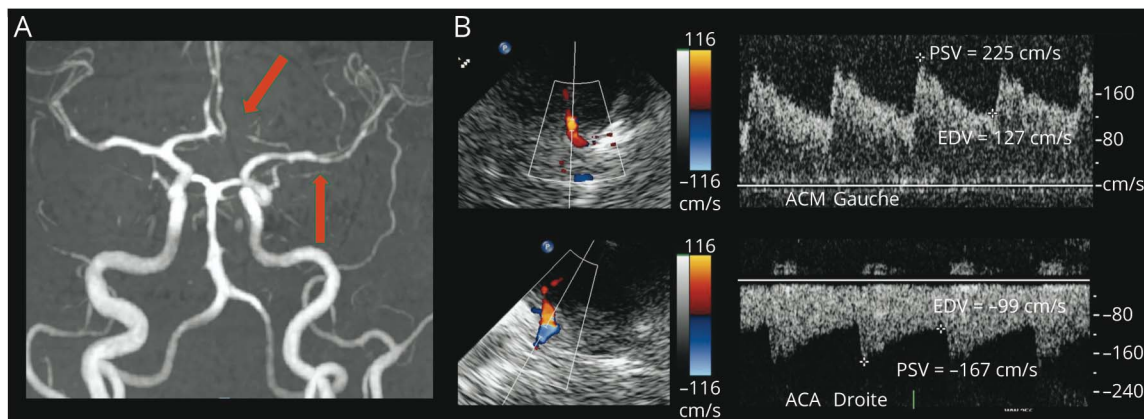
We report the case of a young woman who, in the week following delivery, presented with recurrent headaches leading to the discovery of 3 different and possibly unrelated neurologic complications: postdural puncture headache, followed by CSVT complicated by a venous infarct and asymptomatic RCVS, all of which can present with headaches.

Headache is a frequent symptom in the postpartum period, with a prevalence ranging from 11% to 80%, mostly due to primary headaches.² However, the onset of a new or unusual headache in the peripartum period should be treated as a red

Figure 1 Pre- (A–C) and Post- (D) IVT MRIs



(A) DWI sequence showing a hyperintense signal around the edge of the central sulcus (arrow). (B) TMax perfusion map showing the prolonged time to maximum perfusion in the corresponding area. (C) SWI sequence showing a cortical vein thrombosis (arrow). (D) SWI sequence showing a hemorrhagic transformation. DWI = diffusion-weighted imaging; IVT = IV thrombolysis; SWI = susceptibility-weighted imaging.



(A). Time of flight angiography sequence showing multiple intracranial stenoses (arrows). (B) TCCS (transtemporal approach) color mode (left) with the corresponding Doppler spectrum (right). Upper row: anterior coronal plane, increased PSV and EDV and turbulent flow, corresponding to a left MCA stenosis >50%. Lower row: axial midbrain plane, increased PSV and EDV and turbulent flow, corresponding to a right ACA stenosis of >50%. ACA = anterior cerebral artery; EDV = end-diastolic velocity; MCA = middle cerebral artery; PSV = peak systolic velocity; TCCS = transcranial color-coded duplex ultrasonography.

flag for secondary causes of headaches. This period is associated with an increased risk of several conditions presenting with headaches, including cerebrovascular diseases (subarachnoid hemorrhage, ischemic or hemorrhagic stroke, CSVT, cervical artery dissection, and RCVS) and other causes (posterior reversible encephalopathy syndrome, eclampsia and preeclampsia, PDPH, and pituitary apoplexy).³ On the other hand, the high incidence of headaches in the peripartum period justifies radiologic investigations only in patients with unexpected presentations and using a stepwise approach as in our patient.²

Although PDPH is the most frequent etiology of secondary headaches in the postpartum setting, appearing in up to 5% of pregnancies, other possible etiologies should be kept in mind. Roughly 0.004%–0.01% of pregnancies are complicated by CSVT that causes about 2% of pregnancy-associated strokes.⁴ CSVT occurs mainly in the peripartum period,⁵ probably due to acute changes in coagulation factors and hormones and rapid fluctuations in intracranial pressure during delivery.⁶ CSVT requires acute anticoagulation, which should be initiated despite a hemorrhagic venous infarct as seen in our patient, and the outcome is usually favorable (no or minor disability in 94%).⁷

PDPH and CSVT share common clinical features, which may complicate differential diagnosis. CSVT can present only with headaches, with focal neurologic deficits occurring later or not at all.^{7,8} CSVT consequent to dural puncture during epidural catheter placement has been reported,⁸ although a causal relationship is still unclear.⁹ CSVT has also been described in up to 1%–2% of patients with spontaneous intracranial hypotension. In this context, venous engorgement, traction on cerebral sinuses from brain sagging, and increased blood

viscosity have been proposed as mechanisms favoring CSVT.^{10,11} We could hypothesize similar mechanisms promoting CSVT following dural puncture.

EBP has been suggested as a treatment for CSVT in spontaneous intracranial hypotension¹⁰; therefore, we cannot rule out that the first headache on day 1 postpartum was the result of CSVT and that the 2 blood patches temporarily relieved the symptoms of thrombosis.

Further complicating our patient's case, MRI angiography revealed transient arterial anomalies, and TCD examination exposed blood flow acceleration, indicating RCVS. There are only a few reports of cases of PDPH coexisting with RCVS and CSVT^{12,13} with no obvious causal links between them. Currently, a connection between CSF leakage and cerebral vasospasm is not clearly established, despite a few published cases suggesting intracranial hypotension precipitating vasospasm.¹⁴ If this hypothetical association exists, we could suppose that the intracranial hypotension following the lumbar puncture may have been the primum movens of both the RCVS and CSVT seen in our patient, although further studies are needed to ascertain a causal relationship.

RCVS was formerly known as postpartum angiopathy (PPA) occurring in the 6 weeks following delivery, and its pathogenesis remains unknown. Postpartum RCVS is usually self-limiting with resolution of symptoms and angiographic abnormalities within 3 days to 3 months. However, in severe cases, it can cause ischemic stroke, brain hemorrhage, and/or reversible parenchymal edema. RCVS typically presents with recurrent bouts of sudden extreme headaches, sometimes accompanied by focal neurologic symptoms. If serious,

postpartum RCVS should be treated with nimodipine and perhaps intra-arterial antispastic drugs and balloon angioplasty.¹⁵ In the present case, we considered the RCVS as an incidental finding, as the characteristics of the initial and later headaches seemed better explained by PDPH and CSVT, respectively, and we did not administer any of the treatments because the vasospasms were asymptomatic and relatively mild.

At the initial evaluation in the emergency department, we considered that the patient had an ischemic stroke because of a small cortical diffusion restriction on the MRI (Figure 1) and treated the patient accordingly with IV thrombolysis. However, a proportional hemiparesis as seen in our patient would be expected from a deep territory stroke, such as stroke in the internal capsule or brainstem, and the ischemic lesion in the cortical territory of Figure 1 would rather provoke a focal motor deficit, limited to the cortical representation corresponding to the affected cortical area.

In summary, our patient presented with postpartum headaches initially attributed to and successfully treated as PDPH. The patient then developed hemiparesis followed by a new, contralateral headache from a cortical venous infarct provoked by CSVT, and finally, we found the presence of radiologic (but asymptomatic) RCVS.

Clinicians should therefore be conscious of the possible co-occurrence of multiple etiologies of postpartum headaches and carefully evaluate and correlate all neurologic symptoms using appropriate diagnostic examinations. Early and correct identification of treatable causes of postpartum headaches should help void complications and long-term sequelae.

Acknowledgment

The authors thank Melanie Price Hirt for English language correction of the manuscript.

Study Funding

No targeted funding reported.

Disclosure

The authors report no relevant disclosures. Go to Neurology.org/N for full disclosures.

Publication History

Received by *Neurology* December 31, 2021. Accepted in final form June 30, 2022. Submitted and externally peer reviewed. The handling editor was Roy Strowd III, MD, MEd, MS.

Appendix Authors

Name	Location	Contribution
Leonardo Caranzano, MMed	Department of Clinical Neurosciences, Neurology Service, Lausanne University Hospital and University of Lausanne, Switzerland	Drafting/revision of the manuscript for content, including medical writing for content, and major role in the acquisition of data
Helene Fenter, MMed	Department of Clinical Neurosciences, Neurology Service, Lausanne University Hospital and University of Lausanne, Switzerland	Drafting/revision of the manuscript for content, including medical writing for content, and major role in the acquisition of data
Philippe Maeder, MD, PhD	Department of Medical Radiology, Radiodiagnostic and Interventional Radiology Service, Lausanne University Hospital and University of Lausanne, Switzerland	Drafting/revision of the manuscript for content, including medical writing for content
Patrik Michel, MD, PhD	Department of Clinical Neurosciences, Neurology Service, Lausanne University Hospital and University of Lausanne, Switzerland	Drafting/revision of the manuscript for content, including medical writing for content
Davide Strambo, MD	Department of Clinical Neurosciences, Neurology Service, Lausanne University Hospital and University of Lausanne, Switzerland	Drafting/revision of the manuscript for content, including medical writing for content, and major role in the acquisition of data

References

- Russell R, Laxton C, Lucas DN, Niewiarowski J, Scrutton M, Stocks G. Treatment of obstetric post-dural puncture headache. Part 2: epidural blood patch. *Int J Obstet Anesth*. 2019;38:104-118.
- Stella CL, Jodicke CD, How HY, Harkness UF, Sibai BM. Postpartum headache: is your work-up complete? *Am J Obstet Gynecol*. 2007;196(4):318.e1-318.e7.
- Robbins MS. Headache in pregnancy. *Continuum (Minneapolis, Minn)*. 2018;24(4, Headache):1092-1107.
- James AH, Bushnell CD, Jamison MG, Myers ER. Incidence and risk factors for stroke in pregnancy and the puerperium. *Obstet Gynecol*. 2005;106(3):509-516.
- Jaigobin C, Silver FL. Stroke and pregnancy. *Stroke*. 2000;31(12):2948-2951.
- Miller EC, Leffert L. Stroke in pregnancy: a focused update. *Anesth Analg*. 2020;130(4):1085-1096.
- Kashkoush AI, Ma H, Agarwal N, et al. Cerebral venous sinus thrombosis in pregnancy and puerperium: a pooled, systematic review. *J Clin Neurosci*. 2017;39:9-15.
- Borum SE, Naul LG, McLeskey CH. Postpartum dural venous sinus thrombosis after postdural puncture headache and epidural blood patch. *Anesthesiology*. 1997;86(2):487-490.
- Ghatge S, Uppugonduri S, Kamarzaman Z. Cerebral venous sinus thrombosis following accidental dural puncture and epidural blood patch. *Int J Obstet Anesth*. 2008;17(3):267-270.
- Zhang D, Wang J, Zhang Q, He F, Hu X. Cerebral venous thrombosis in spontaneous intracranial hypotension: a report on 4 cases and a review of the literature. *Headache*. 2018;58(8):1244-1255.
- Pensato U, Giammello F, Baldini T, et al. The domino effect of cephalgic spontaneous intracranial hypotension. *Neurol Sci*. 2021;42(1):309-312.
- Hoeren M, Hader C, Strümpell S, Weiller C, Reinhard M. Peripartum angiopathy with simultaneous sinus venous thrombosis, cervical artery dissection and cerebral arterial vasoconstriction. *J Neurol*. 2011;258(11):2080-2082.
- Katzin LW, Levine M, Singhal AB. Dural puncture headache, postpartum angiopathy, pre-eclampsia and cortical vein thrombosis after an uncomplicated pregnancy. *Cephalalgia*. 2007;27(5):461-464.
- Chaves C, Freidberg SR, Lee G, Zerris V, Ries S, Chavali R. Cerebral vasospasm following intracranial hypotension caused by cerebrospinal fluid leak from an incidental lumbar durotomy. Case report. *J Neurosurg*. 2005;102(1):152-155.
- Singhal AB, Bernstein RA. Postpartum angiopathy and other cerebral vasoconstriction syndromes. *Neurocrit Care*. 2005;3(1):91-97.

Neurology[®]

Pearls & Oy-sters: Postdural Puncture Headache, Cerebral Sinus Venous Thrombosis, and Reversible Cerebral Vasoconstriction Syndrome in the Peripartum Period

Leonardo Caranzano, Helene Fenter, Philippe Maeder, et al.

Neurology 2022;99;665-668 Published Online before print August 10, 2022

DOI 10.1212/WNL.0000000000201111

This information is current as of August 10, 2022

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/99/15/665.full
References	This article cites 15 articles, 1 of which you can access for free at: http://n.neurology.org/content/99/15/665.full#ref-list-1
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): Cerebral venous thrombosis http://n.neurology.org/cgi/collection/cerebral_venous_thrombosis Secondary headache disorders http://n.neurology.org/cgi/collection/secondary_headache_disorders Stroke in young adults http://n.neurology.org/cgi/collection/stroke_in_young_adults
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2022 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

