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## Case Report

# MRA and ASL perfusion findings in pediatric reversible cerebral vasoconstriction syndrome

Samar Kayfan, BA, Arghavan Sharifi, BS, Sophia Xie, MD, Chen Yin, MD, Cory M. Pfeifer, MD, MS\*

Department of Radiology, University of Texas Southwestern Medical Center, 5323 Harry Hines Blvd, Dallas, TX 75390, USA

## ARTICLE INFO

## Article history:

Received 1 March 2019

Revised 7 April 2019

Accepted 7 April 2019

Available online 28 April 2019

## Keywords:

Reversible cerebral vasoconstriction syndrome

## ABSTRACT

Reversible vasoconstriction is rare in the pediatric population. Typically manifesting as peripheral vasoconstriction with variable neurologic symptoms, Reversible cerebral vasoconstriction syndrome is often a diagnosis of exclusion and may not be diagnosed in the absence of angiographic imaging. Shown here are 2 cases of pediatric Reversible cerebral vasoconstriction syndrome with disparate MRI findings and arterial spin labeling perfusion findings.

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## Introduction

Reversible cerebral vasoconstriction syndrome (RCVS) is a clinico-radiological condition that presents with recurrent severe headaches with or without concurrent focal neurologic signs. Seizures have been described. Calabrese et al established diagnostic criteria for this syndrome in 2007 [1,2]. Most reported cases of RCVS have been in adult patients with predilection for female gender. Some possible triggers of RCVS include pregnancy, the use of serotonin agonist medications, and illicit drug use [3,4]. Most patients initially present with severe headaches, sometimes characterized as “thunderclap”

headaches. Imaging findings described in the reported adult cases include multifocal areas of vasoconstriction in the cerebral arteries which are transient in nature.

It is important to rule out other diagnoses including venous sinus thrombosis, pituitary apoplexy, cervical artery dissection, and intraparenchymal hemorrhages [5–7], as these disorders may produce similar headaches. Posterior reversible encephalopathy can share clinical features with RCVS and may share a similar pathophysiology [3,8].

In contrast to the adult population, RCVS in children is not well-understood with few reported cases in medical literature. Presented here are 2 pediatric cases of RCVS along with their imaging findings and clinical courses.

Funding: This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

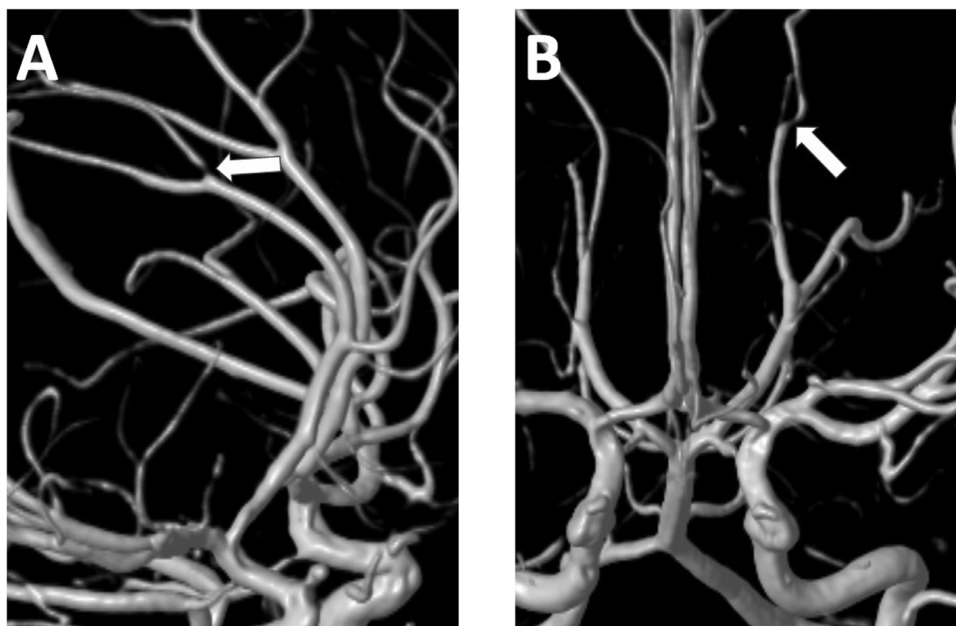
Conflict of interest: The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

\* Corresponding author.

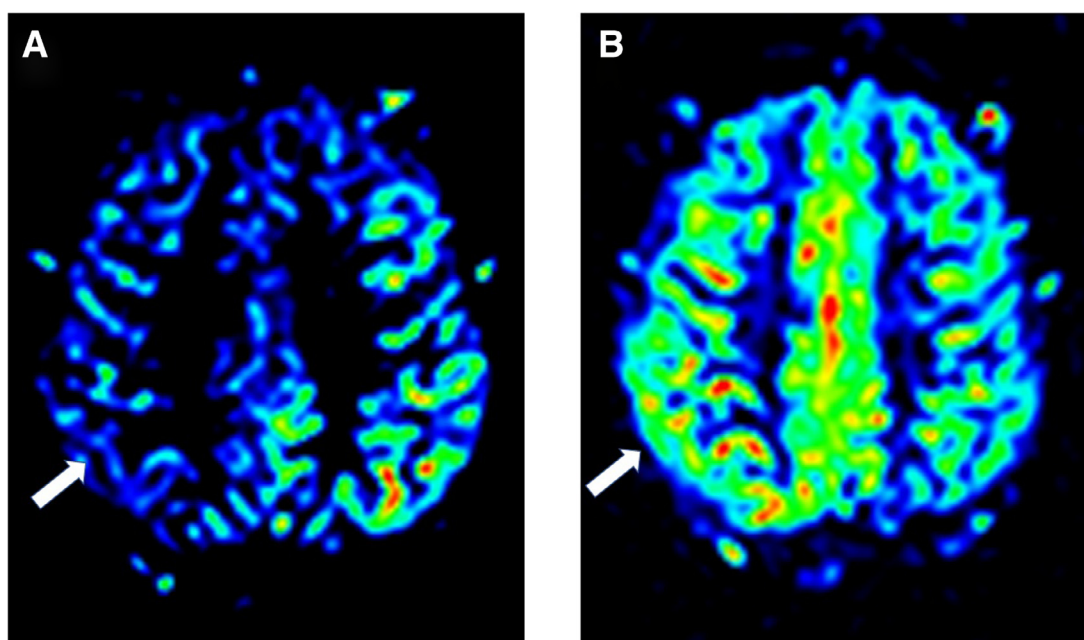
E-mail address: [Cory.Pfeifer@utsouthwestern.edu](mailto:Cory.Pfeifer@utsouthwestern.edu) (C.M. Pfeifer).

<https://doi.org/10.1016/j.radcr.2019.04.010>

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**Fig. 1** – Three-dimensional reconstructions of an MRA of the brain showing focal narrowing of a peripheral branch of the right anterior cerebral artery (A) and left parieto-occipital artery (B).

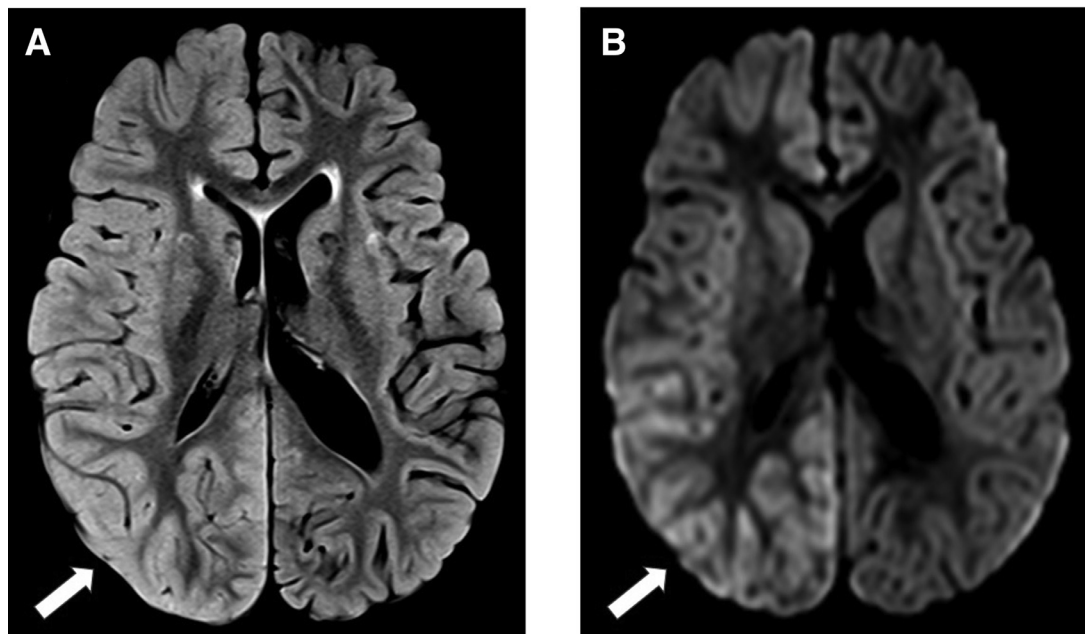


**Fig. 2** – ASL perfusion map performed the day following admission (A) reveals decreased perfusion in the right cerebral hemisphere. Follow-up study obtained 10 months later (B) demonstrates relative hyperperfusion of the right hemisphere.

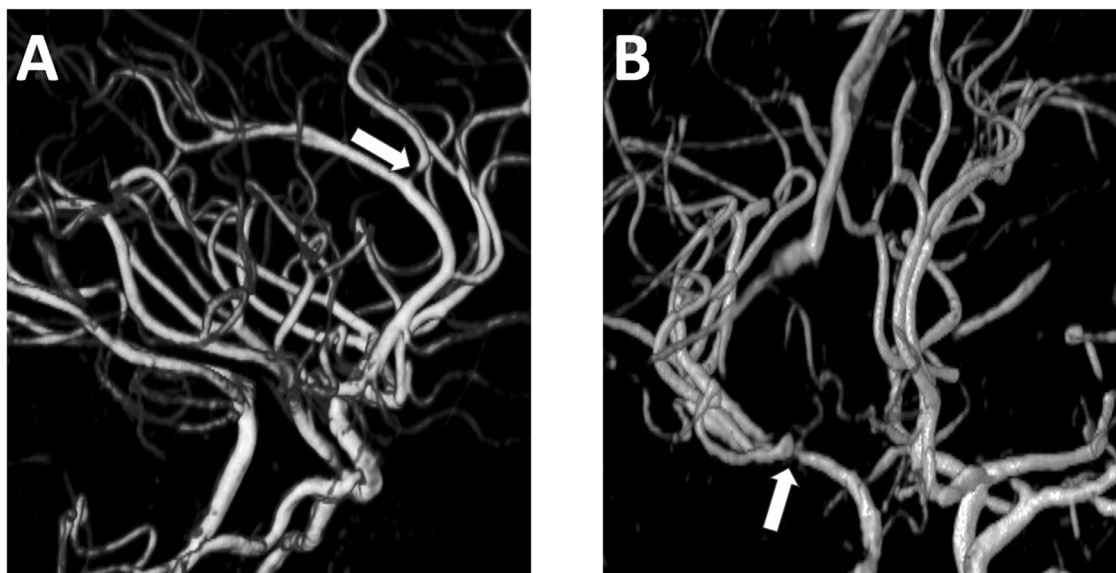
### Case report 1

The first patient is a 13-year-old female with a history of headaches who initially presented to the emergency department with throbbing headache, altered mental status, confusion, and slurred speech. The patient localized her headache to left and frontal regions and reported nausea. She described the pain as 8/10. Shortly after the initial onset of cephalgia, she

experienced blurred vision in her right eye. She then slept 2 hours. When she awoke, her headache had increased in severity, and her vision had not improved. The patient then began to have slurred and incomprehensible speech. She was confused and not oriented to time, though she was oriented to person and place. Throughout this time, the patient did not experience any frank tonic-clonic activity, eye deviation, spontaneous voiding/defecating, or loss of consciousness. She has no family history of migraines, headaches, hypertension, strokes,



**Fig. 3 – Axial FLAIR (A) exhibits cortical T2 hyperintense signal throughout the right cerebral hemisphere with associated diffusion restriction (B).**



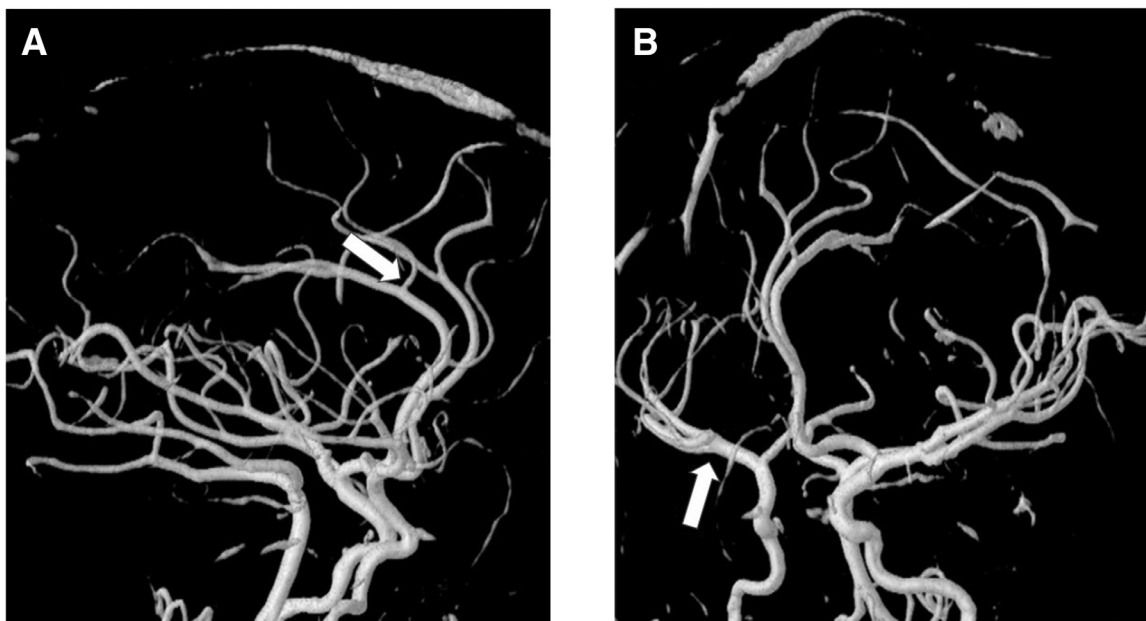
**Fig. 4 – Three-dimensional reconstructions of an MRA of the brain showing focal narrowing of a peripheral branch of the right anterior cerebral artery (A) and right middle cerebral artery at the bifurcation (B).**

or seizures. CT of the head performed at presentation demonstrated no abnormality. Her complete-blood count, comprehensive metabolic profile, and urine-drug screening results were all unremarkable. She received solumedrol, compazine, zofran, toradol, and a normal-saline bolus.

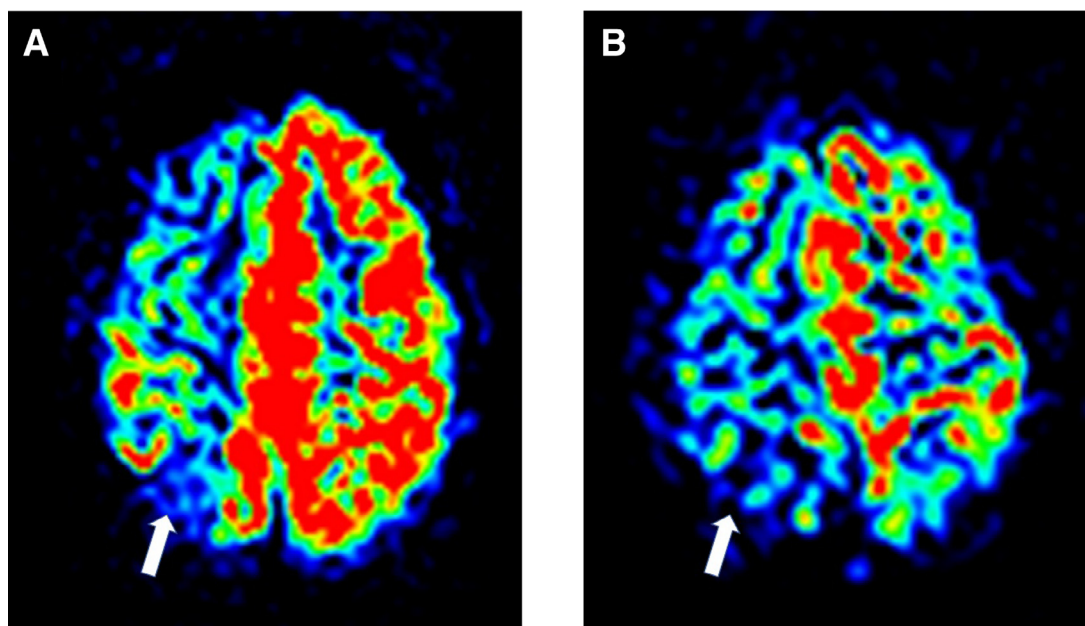
On the second day of admission, the patient underwent Magnetic Resonance Imaging (MRI) which showed no focal parenchymal signal abnormality. There was no diffusion restriction. Concomitant Magnetic Resonance Angiogram (MRA) of the brain demonstrated multiple areas of narrowing of several peripheral branches of the Circle of Willis includ-

ing the right anterior cerebral artery (Fig. 1A) as well as the left parieto-occipital artery (Fig. 1B). An arterial spin labeling (ASL) perfusion study showed decrease in cerebral blood flow throughout the right cerebral hemisphere (Fig. 2A).

After 3 months of verapamil therapy, the patient only reported 2 headaches which were less intense and shorter in duration than her presenting symptoms. The patient denied altered mental status, confusion, and slurred speech. Repeat MRA of the head performed 10 months following initial presentation showed an improved appearance of the multiple arterial stenoses. There was asymmetric hyperperfusion of the



**Fig. 5** – Three-dimensional reconstructions of an MRA of the brain now show improvement in narrowing of a peripheral branch of the right anterior cerebral artery (A) and right middle cerebral artery at the bifurcation (B).



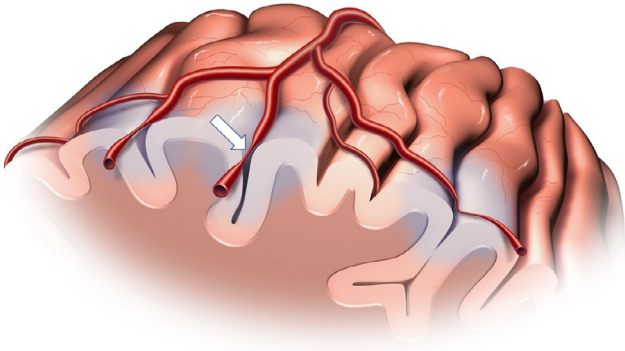
**Fig. 6** – ASL perfusion map reveals decreased perfusion throughout the right cerebral hemisphere (A). Follow-up study obtained the following month (B) exhibits continued decreased perfusion in the right hemisphere.

right cerebral hemisphere compared to the left (Fig. 3). There was no focal parenchymal signal abnormality in the brain or diffusion restriction.

### Case report 2

The second patient is a 6-year-old male who presented with headache, slurred speech, and left-sided weakness. His past medical history was significant for recurrent headaches and

transient right-sided weakness. CT of the head and MRI of the brain performed 7 months prior were unremarkable. MRI of the brain ordered at this presentation was significant for cortically based increased FLAIR signal throughout the right cerebral hemisphere (Fig. 3A) with associated diffusion restriction (Fig. 3B). MRA demonstrated focal narrowing of a distal branch of the right anterior cerebral artery (Fig. 4A) and right middle cerebral artery at the bifurcation (Fig. 4B). There was decreased ASL perfusion throughout the right cerebral hemisphere (Fig. 6A).



**Fig. 7 – Illustration depicts a focally narrowed distal cerebral artery (arrow) with underlying decreased perfusion.**

After 1 month of verapamil therapy, repeat MRI and MRA showed improvement in vasoconstriction (Fig. 5) with improvement in ASL perfusion (Fig. 6B).

## Discussion

Coffino and Fryer [3] reviewed the 13 cases of pediatric RCVS that had been reported in the literature as of 2017. Of these 13 children, 11 were male. This represents a striking reversal of sex predominance relative to the adult population in which the female portion of RCVS cases has been reported to be as high as 81% [4].

These 2 cases highlight the unique radiological presentation of RCVS. In the first case, there were no parenchymal imaging findings on MRI despite the stenoses present on the MRA. In the second case, there was diffusion restriction throughout the affected hemisphere accompanying the MRA findings. MRI can show variable findings in the same patient from presentation to presentation. Indeed, review of the second patient's medical record showed prior episodes of headache and transient weakness in the setting of a normal MRI.

Little is known of the relationship between ASL perfusion and RCVS in the pediatric patient. The finding of slightly increased perfusion 10 months following initial presentation in the first patient may suggest a reperfusion hyperemia in

response to calcium channel blocker therapy. In the second patient, the follow-up ASL perfusion study may have been performed too recently to observe a similar phenomenon.

RCVS is a transient vasculopathy which can present as a severe headache with distinguishing features on MRA studies of multifocal segment cerebral artery vasoconstriction. Areas of narrowing are typically observed more peripherally (Fig. 7). Focal stenosis of a peripheral artery with subsequent parenchymal hypoperfusion is depicted in the second patient. Calcium channel blocker therapy has been used to improve clinical symptomatology and vasoconstriction.

RCVS is likely underdiagnosed in the pediatric population but should be considered among patients who present with recurrent thunderclap or severe headaches. As such, when neurologic imaging is considered in the approach to headache, MRA and/or ASL perfusion may be important adjuncts to conventional MRI of the brain.

## REFERENCES

- [1] Calabrese LH, Dodick DW, Schwedt TJ, Singhal AB. Narrative review: reversible cerebral vasoconstriction syndromes. *Ann Intern Med* 2007;146:34–44.
- [2] Call GK, Fleming MC, Sealfon S, Levine H, Kistler JP, Fisher CM. Reversible cerebral segmental vasoconstriction. *Stroke* 1988;19(9):1159–70.
- [3] Coffino SW, Fryer RH. Reversible cerebral vasoconstriction syndrome in pediatrics: a case series and review. *J Child Neurol* 2017;32:614–23.
- [4] Singhal AB, Hajj-Ali RA, Topcuoglu MA, Fok J, Bena J, Yang D, et al. Reversible cerebral vasoconstriction syndromes: analysis of 139 cases. *Arch Neurol* 2011;68:1005–12.
- [5] Kamide T, Tsutsui T, Misaki K, Sano H, Mohri M, Uchiyama N, et al. A pediatric case of reversible cerebral vasoconstriction syndrome with similar radiographic findings to posterior reversible encephalopathy syndrome. *Pediatr Neurol* 2017;71:73–6.
- [6] Kazato Y, Fujii K, Oba H, Hino M, Ochiai H, Uchikawa H, et al. Reversible cerebral vasoconstriction syndrome associated with brain parenchymal hemorrhage. *Brain Dev* 2012;34:696–9.
- [7] Yoshioka S, Takano T, Ryuji F, Takeuchi Y. A pediatric case of reversible cerebral vasoconstriction syndrome with cortical subarachnoid hemorrhage. *Brain Dev* 2012;34:796–8.
- [8] Ducros A. Reversible cerebral vasoconstriction syndrome. *Lancet Neurol* 2012;11:906–17.