Pediatric Reversible Cerebral Vasoconstriction Syndrome: A Case Requiring Operative Intervention and Literature Review

Gabriel Pham, M.S.1, Michael Robinson, M.D. Ph.D.1, Michael Taylor, M.D.2, Katrina Peariso, M.D. Ph.D.2, Todd Abruzzo, M.D.1, and Sudhaker Vadivelu, D.O.3

1Division of Pediatric Neurosurgery, Cincinnati Children's Hospital Medical Center, Cincinnati, OH. 2Division of Child Neurology, Cincinnati Children's Hospital Medical Center, Cincinnati, OH. 3Department of Radiology, Phoenix Children's Medical Group, Phoenix, AZ.

Introduction
Reversible cerebral vasoconstriction syndrome (RCVS) is a clinical-radiological syndrome characterized by (1,2,5):
• bilateral, diffuse, segmental narrowing of cerebral arteries with “string of beads” appearance
• Spontaneous resolution of vasoconstriction several weeks after acute episode
• Treatment is largely supportive with good prognosis.

Pediatric RCVS is likely underdiagnosed and poorly reported in case literature (3,4). We report a 9 year old with angiographically-confirmed RCVS and review the literature review of reported cases.

Methods
Clinical data and imaging were abstracted for the index patient’s medical record. The patient has been followed serially for 3 years in our pediatric Cerebrovascular Center.

Criteria
Recurrent thunderclap headaches with/without neurologic sx
Uniphasic disease course with no recurrence
No evidence of aneurysmal SAH
Normal or near normal CSF findings
Multifocal segmental cerebral artery vasoconstriction
Reversibility of angiographic abnormalities within 12 weeks

Table 1 Diagnostic Criteria for RCVS

Case
Patient is a 9 year old female chronically on stimulant therapy for ADHD who developed symptoms after co-administration of sympathomimetic cold medicine. She presented to the ED with 4 days of thunderclap headaches, irritability, emesis, and encephalopathy. Imaging obtained at admission is shown in Figure 1 showing intracranial hemorrhage, obstructive hydrocephalus, vasoconstriction, and angiography demonstrating characteristic “beading” pattern. Patient was treated with CSF diversion with external ventricular drain, clot removal, and medical management of increased ICP and seizures (Figure 2). Patient also underwent angiography with intra-arterial verapamil x2 demonstrating transient arterial vasodilation. 3 month follow-up imaging shows resolution of cerebral vasoconstriction.

Table 2 Aggregated Demographic and Clinical Features of Reported Cases of Pediatric Reversible Cerebral Vasocclusion. RCVS in the pediatric population shows a male predominance. The majority of patients will present with headache. Seizures was the most common neurologic symptom. Females are more likely have neurologic deficits and seizures. SAH in adolescent RCVS patients are female while SAH in children are male. Parenchymal hemorrhage only has been documented in female pediatric patients. *A case report of a 4 month old did not include patient’s gender.

Conclusions
Important aspects of our case
• Complications from RCVS in may warrant surgical treatment
• Intra-arterial verapamil transiently alleviates vasoconstriction and symptoms, but may not change clinical course
• Treatment with anitconvulsants for seizure prophylaxis and calcium channel blockers for blood pressure management may be beneficial

Summary of Findings for Pediatric RCVS from Literature

Table 2. Aggregated Demographic and Clinical Features of Reported Cases of Pediatric Reversible Cerebral Vasocclusion. RCVS in the pediatric population shows a male predominance. The majority of patients will present with headache. Seizures was the most common neurologic symptom. Females are more likely have neurologic deficits and seizures. SAH in adolescent RCVS patients are female while SAH in children are male. Parenchymal hemorrhage only has been documented in female pediatric patients. *A case report of a 4 month old did not include patient’s gender.

Works Cited and Acknowledgements