

Reversible Cerebral Vasoconstriction Syndrome

- a.k.a. Call-Fleming syndrome
- Group of disorders characterized by
- Reversible, multifocal cerebral artery vasoconstrictions
- Severe headaches \pm focal neurological deficits

General Features

● Etiology

- Thought to represent transient disturbance in control of cerebral vascular tone → vasoconstriction → ischemia, stroke, death

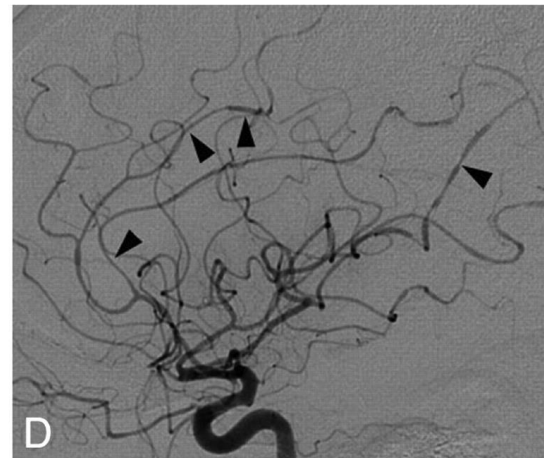
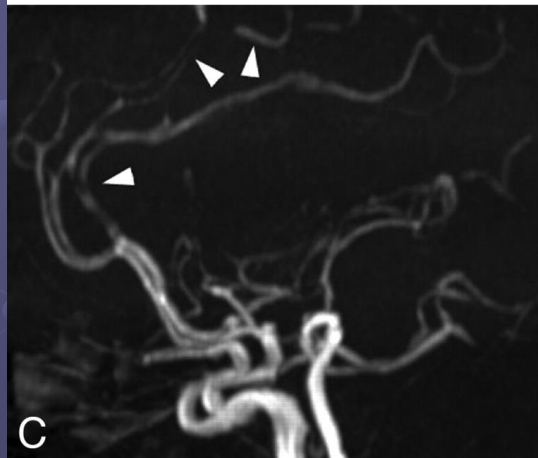
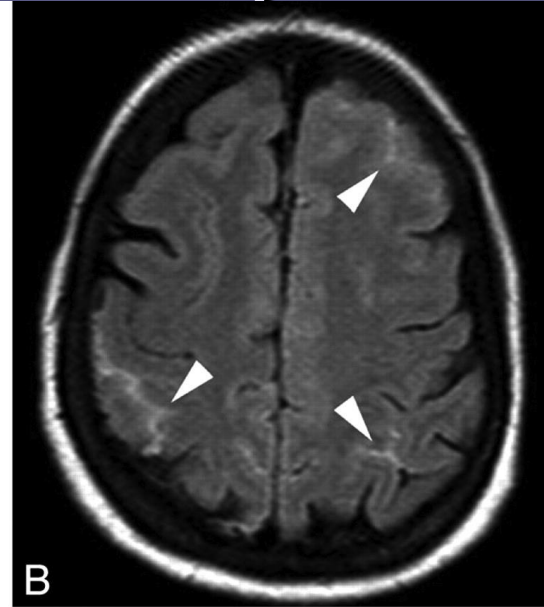
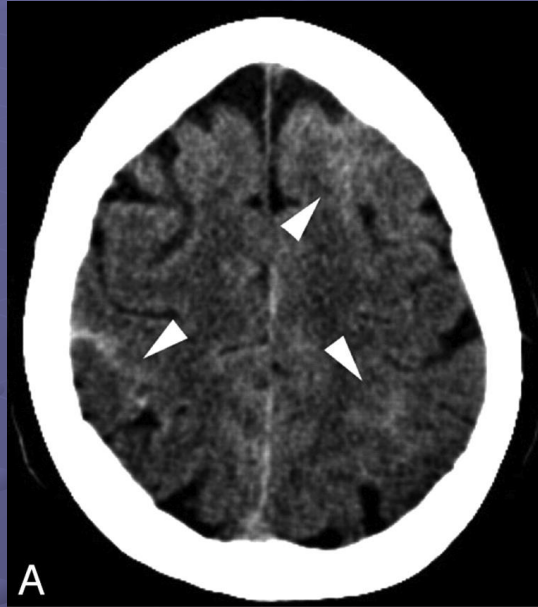
● Associated abnormalities

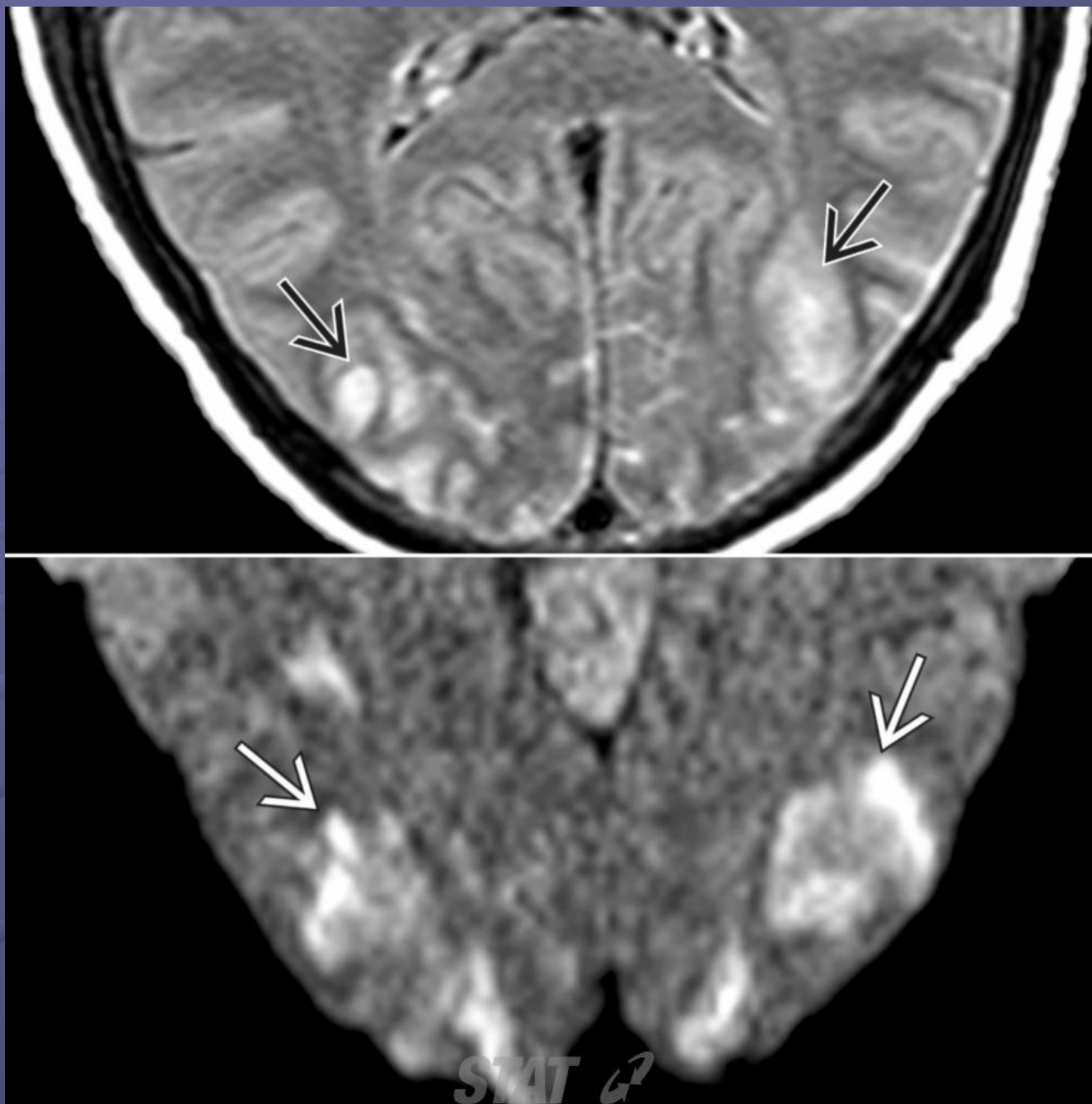
- Occurs spontaneously (1/3 of cases) or may be precipitated by
 - Postpartum state
 - Exposure to vasoactive substances
 - Cannabis, cocaine, ecstasy, amphetamine derivatives, LSD
 - Selective serotonin reuptake inhibitors (SSRIs)
 - Nasal decongestants, pseudoephedrine
 - Ergotamine tartrate, bromocriptine, sumatriptan
 - Pheochromocytoma, bronchial carcinoid tumor

Imaging

- Acute/recurrent headaches with vasculitic pattern (DSA)
- DSA = crucial for diagnosis (100% sensitive)
 - Involves large, medium-sized arteries
 - Diffuse, multifocal, segmental narrowing
 - Sometimes "string of beads" or "sausage strings"
- NECT often negative
 - Small cortical subarachnoid hemorrhage (SAHs) (20%) \pm parenchymal hemorrhage
- CTA/MRA: May be normal if subtle changes (10%)
 - Diffuse segmental arterial constriction in 90%
 - May require DSA for diagnosis (100% sensitive)
 - Interval DSA may show improvement with vasodilator Rx
- TCD: \uparrow arterial velocities in MCA, ICA, ACA

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MR obtained 4 days after admission in the same patient when she noticed visual disturbances. FLAIR scan (top) shows multifocal hyperintensities in both occipital lobes (black solid arrow). DWI (bottom) shows restricted diffusion (white solid arrow), consistent with ischemia or infarction.