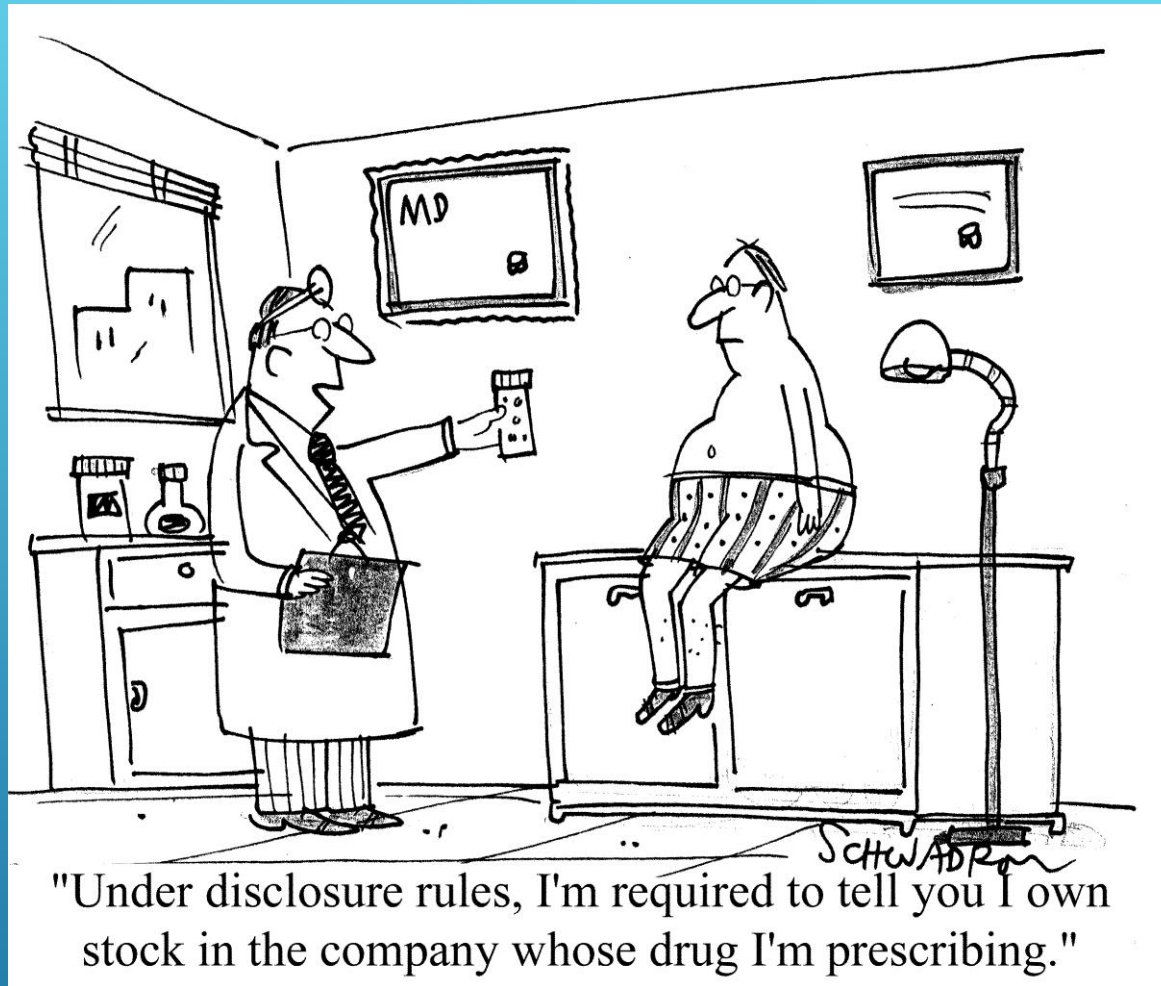


CEREBRAL VENOUS SINUS THROMBOSIS (CVST) AND RELEVANT HYPERCOAGULABLE STATES

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Stroke Medical Director

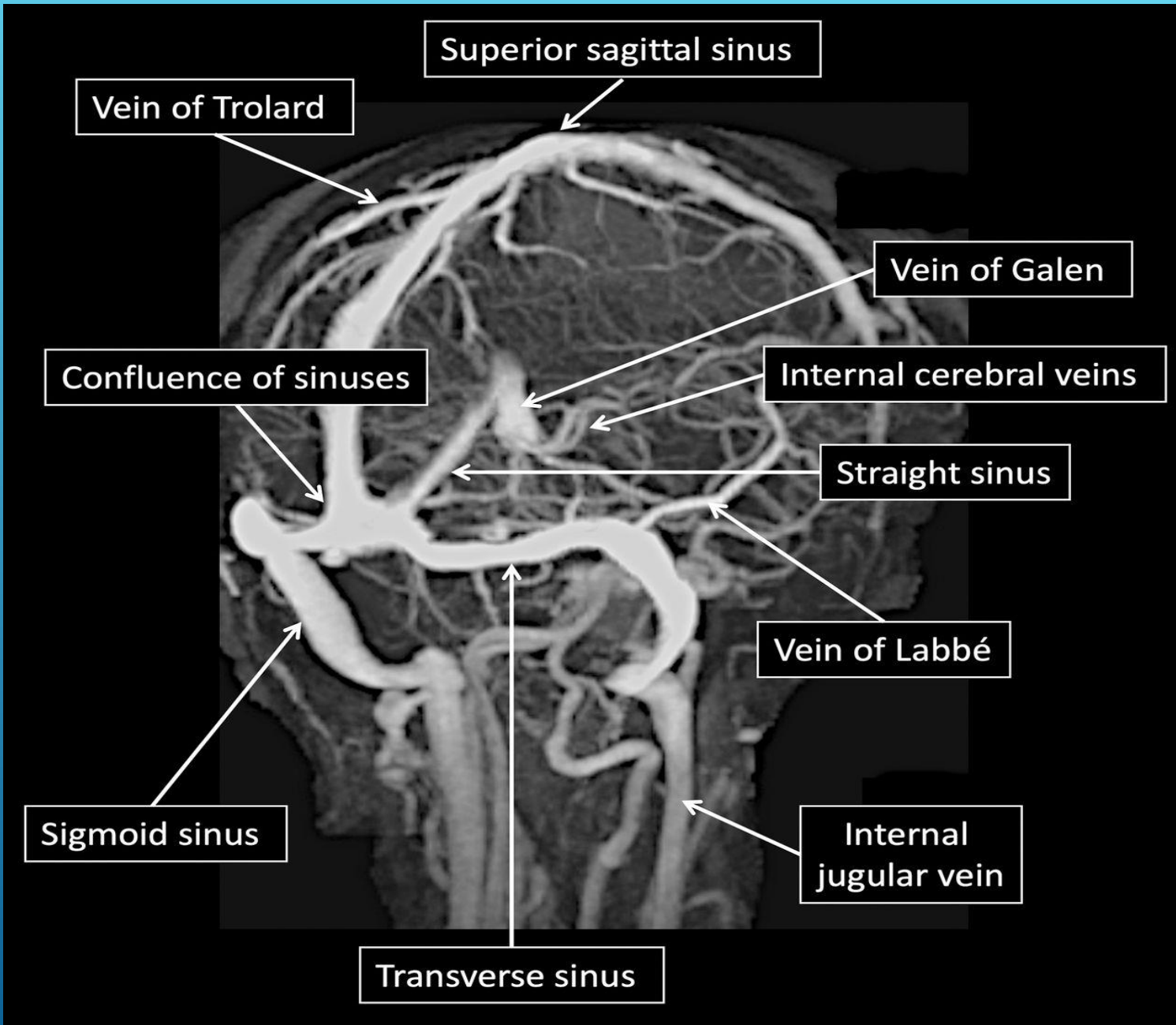
Saint Luke's Health System



NO DISCLOSURES

- ▶ Thrombus in the dural venous sinuses (CVST) or the cortical veins (CVT)
- ▶ Can be complete or partial occlusion(s)
- ▶ Can affect multiple venous sinuses or a mix of the major cerebral venous sinuses and the smaller feeding cortical veins
- ▶ Blood may leak into the parenchyma causing ICH
- ▶ Infrequently, CSF absorption/flow is blocked and hydrocephalus can occur

WHAT IS CVST?



Anatomy of the major intracerebral veins and dural venous sinuses

Ulivi, L. et al. (2020) Cerebral venous thrombosis: A practical guide, *Practical Neurology*. BMJ Publishing Group Ltd. Available at: <https://pn.bmj.com/content/20/5/356> (Accessed: November 3, 2022).

- ▶ 3x as common in women vs. men in younger age groups (20-35)
 - ▶ Likely due to an increased incidence with pregnancy, the puerperium and with oral contraceptive use
- ▶ Equally prevalent between the sexes after the age of 55
 - ▶ Malignancy can be a factor in up to 25% of patients older than 55
- ▶ Affects about 5 people in 1 million each year

STATISTICS

- ▶ Hemorrhagic infarction occurs in 10-50% of cases
- ▶ The superior sagittal sinus is affected in about 62% of cases and the transverse sinus(es) in about 45%
- ▶ 1/3 of cases have more than one sinus affected
- ▶ 30-40% have BOTH sinus(es) and cortical vein(s) affected

MORE STATISTICS

A series of white diagonal lines of varying lengths and thicknesses, located in the bottom right corner of the slide.

- ▶ OCP use/Pregnancy/puerperium
- ▶ Thrombophilias—factor V Leiden, prothrombin gene mutation, antithrombin, protein C and S deficiencies, antiphospholipid antibody syndrome, sickle cell anemia
- ▶ Infections of the head and neck
- ▶ Malignancy
- ▶ Connective tissue disorders, collagen vascular diseases
granulomatous disorders, inflammatory bowel disease
- ▶ Direct injury to the head or neck
- ▶ post-vaccination instances and pro-inflammatory infections such as COVID 19

RISK FACTORS FOR DEVELOPMENT OF CVST (ADULTS)

- ▶ Headache is the most common (70-90%)
- ▶ Focal deficits such as hemiparesis, hemisensory deficits, aphasia
- ▶ Seizures
- ▶ Impairment of consciousness, vomiting
 - ▶ Often if deep cerebral veins are involved affecting the bilateral thalami
- ▶ Papilledema/vision changes (can resemble IIH)
- ▶ With cavernous sinus thrombosis, chemosis, proptosis and painful ophthalmoplegia can occur



PRESENTING SIGNS/SYMPTOMS

- ▶ Often can present insidiously, as clot forms slowly, and collaterals are often present
- ▶ Headache and seizures can differentiate this from AIS, as well as an infarction that is not in a classic single arterial territory, often with a hemorrhagic component
- ▶ Rarely, hydrocephalus can occur as impaired CSF flow
- ▶ Thrombocytopenia—vaccine-induced immune thrombotic thrombocytopenia (VITT)

OTHER CONSIDERATIONS

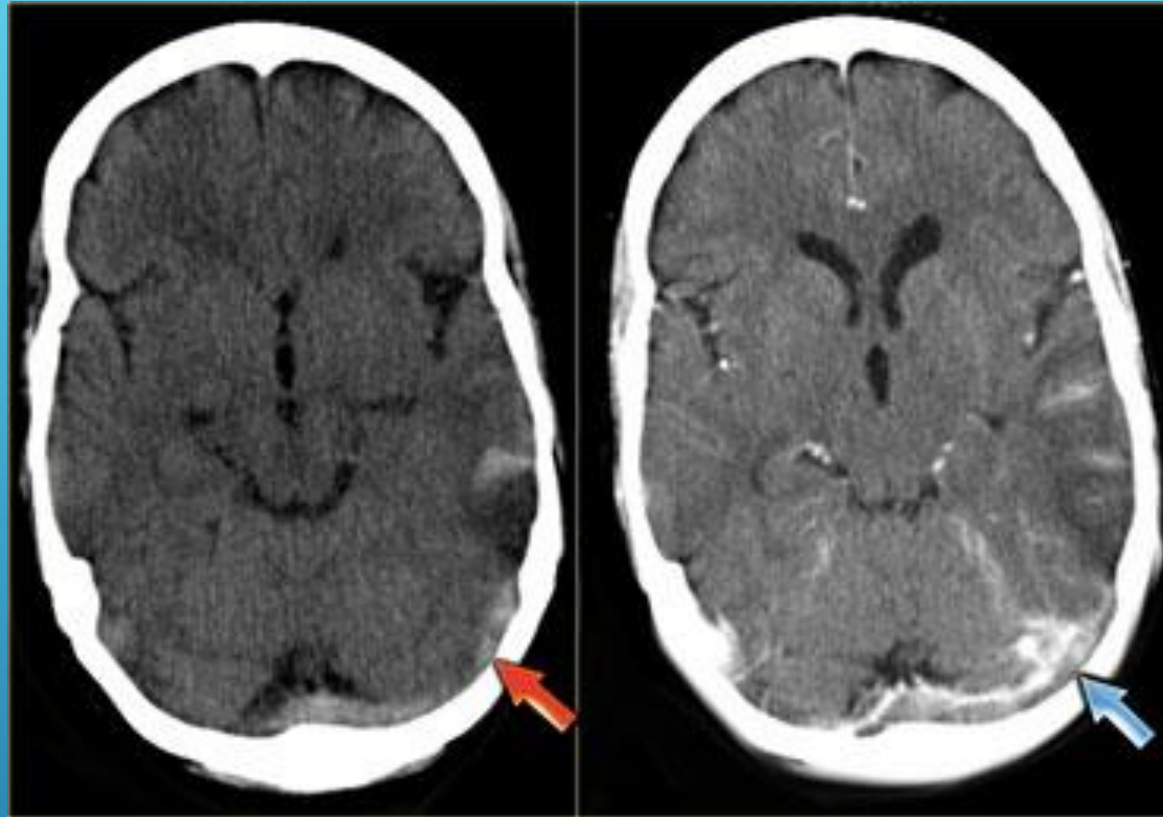
- ▶ Delta sign on contrasted CT head
 - ▶ Contrast outlines thrombus in the torcular Heterophili (confluences of the sinuses)
- ▶ MRI with MRV
 - ▶ Most sensitive for involvement of the cortical veins
- ▶ CT Venogram
- ▶ Cerebral Angiography—test of choice to distinguish between congenital hypoplasia vs thrombus



DIAGNOSIS

- ▶ Pregnancy testing
- ▶ Hypercoagulable studies—may need to be repeated in 12 weeks or after patient is off AC
- ▶ CBC, CMP
- ▶ COVID testing
- ▶ ESR/CRP/D-dimer
- ▶ Consideration for malignancy
- ▶ Testing for connective tissue disorders

OTHER DIAGNOSTIC CONSIDERATIONS



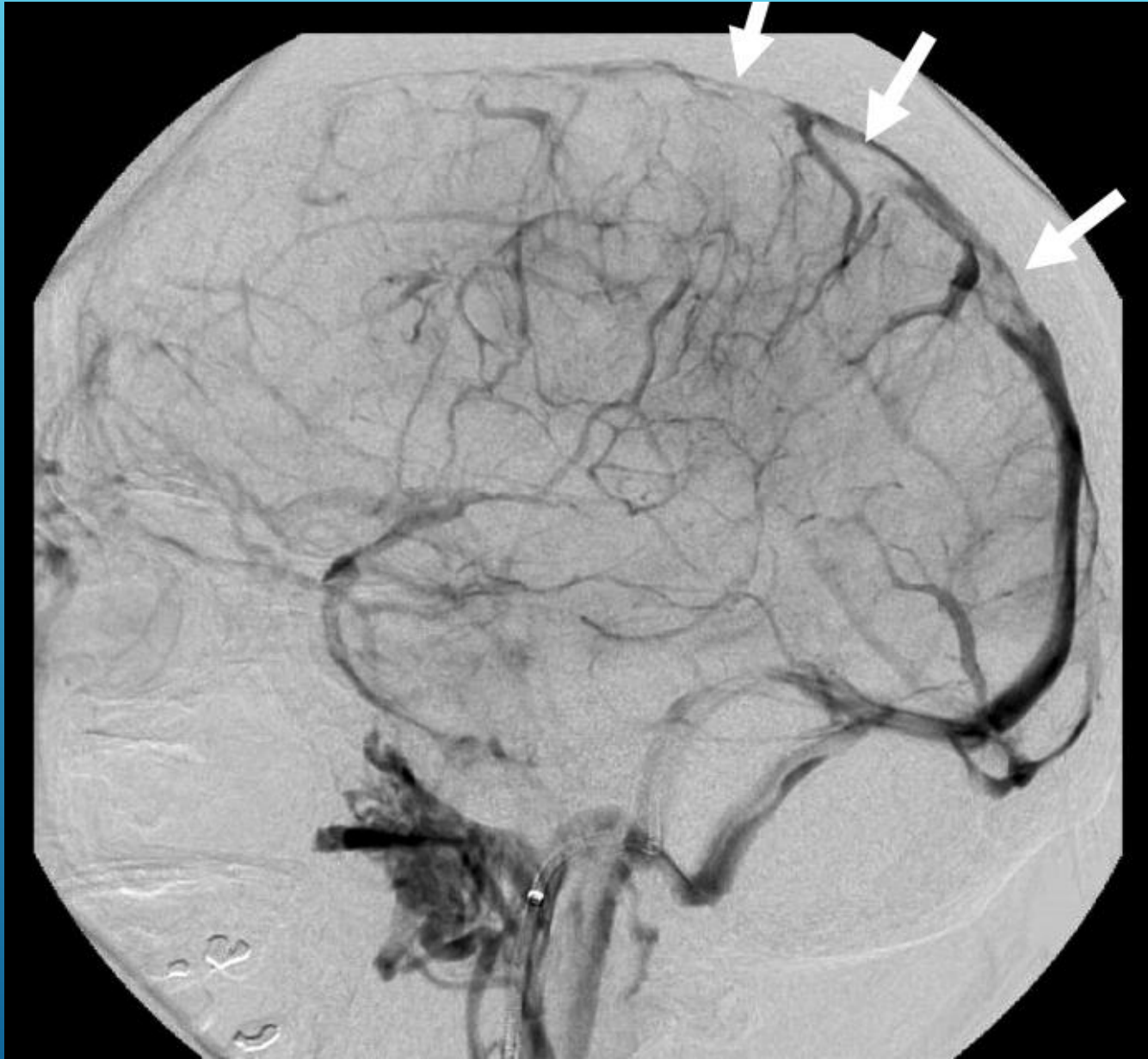
NCCT head with hyperdense acute thrombus in the left transverse sinus and deep cortical veins with associated hemorrhage and hypodensity representing venous infarct in the left temporal lobe



NCCT head with hypodensity (infarcts) in the bilateral thalami and thrombus in the deep cerebral veins

Left transverse CVST
on MRV





Cerebral
angiography with
Superior Sagittal
Sinus thrombosis

- ▶ The mainstay of treatment is heparin drip with transition to oral anticoagulant
- ▶ DOACs vs Warfarin studied in ACTION-CVT—associated with similar clinical and radiographic outcomes and favorable safety profile
- ▶ Occasionally, various angiographic treatments are necessary
- ▶ Treatment in provoked cases can be anticoagulated 3-6 months, but up to 12% recur and up to 14% have a different form of DVT
- ▶ 57-86% have complete functional recovery; 5.5-18% mortality
- ▶ Some persons require AEDs

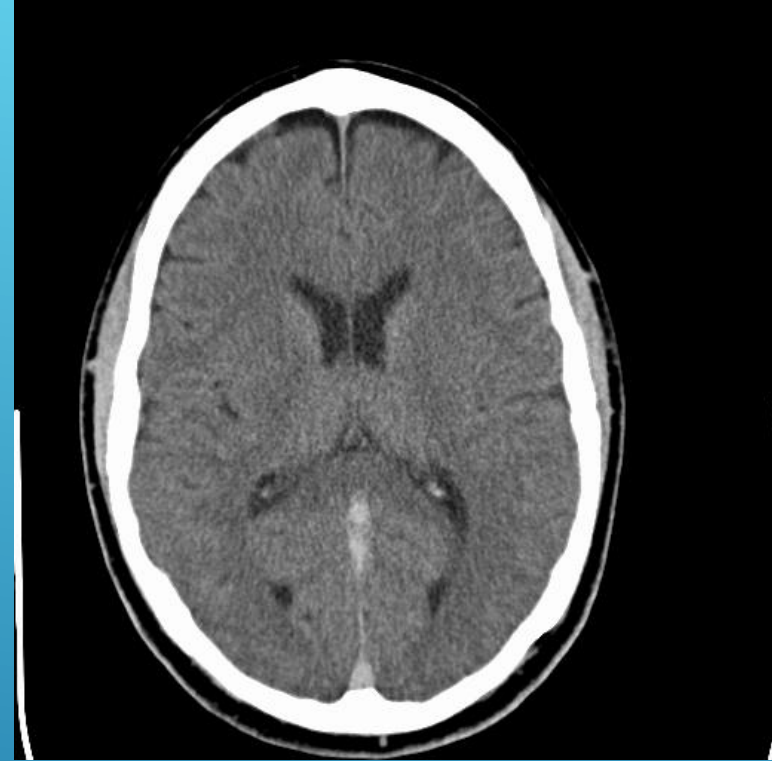
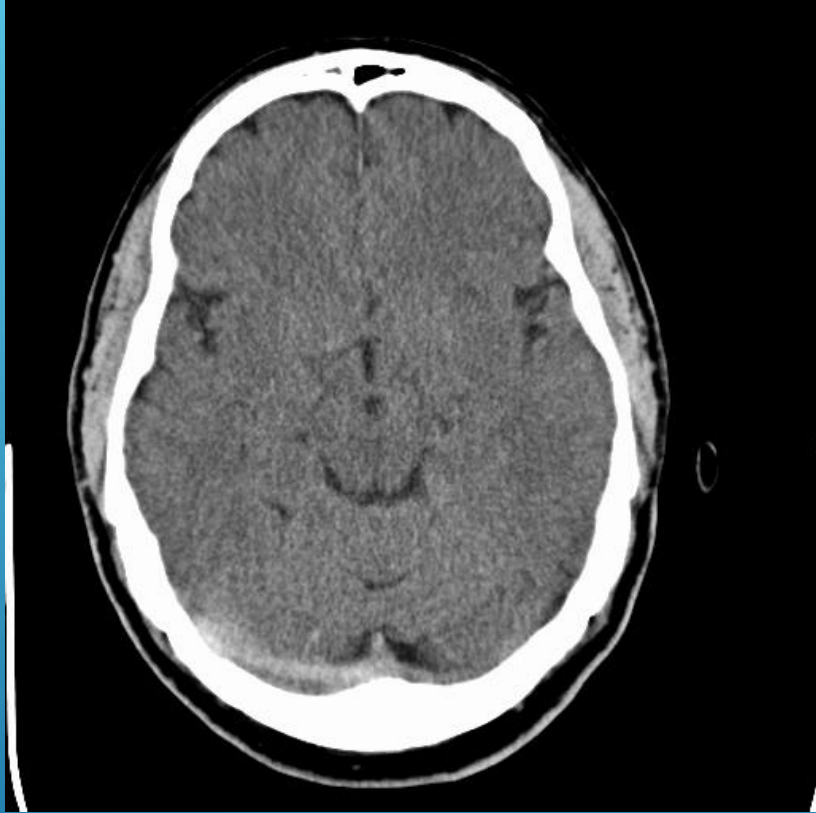
TREATMENT AND PROGNOSIS

- ▶ One study found the CVST incidence rate in the inpatient COVID+ population before vaccines were widely used was 231 per 1,000,000 person-years.
 - ▶ Older patients were more likely to die (45.5% over the median age of 46.5 years, 0% under the median)
 - ▶ Males were more likely to die (44.4% vs 7.7% females)
 - ▶ Those treated with inpatient steroids were more likely to die (44.4% vs 7.7%)
- ▶ CVST has also been seen in higher rates in the population of patients receiving the COVID vaccines that use an adenoviral vector, AstraZeneca and Janssen vaccines
 - ▶ These cases were more likely to be associated with thrombocytopenia, termed CVST with vaccine-induced thrombotic thrombocytopenia
 - ▶ Optimal treatment unclear, but avoid heparin—consider DOAC, other anticoagulants, IVIG, and some propose steroids

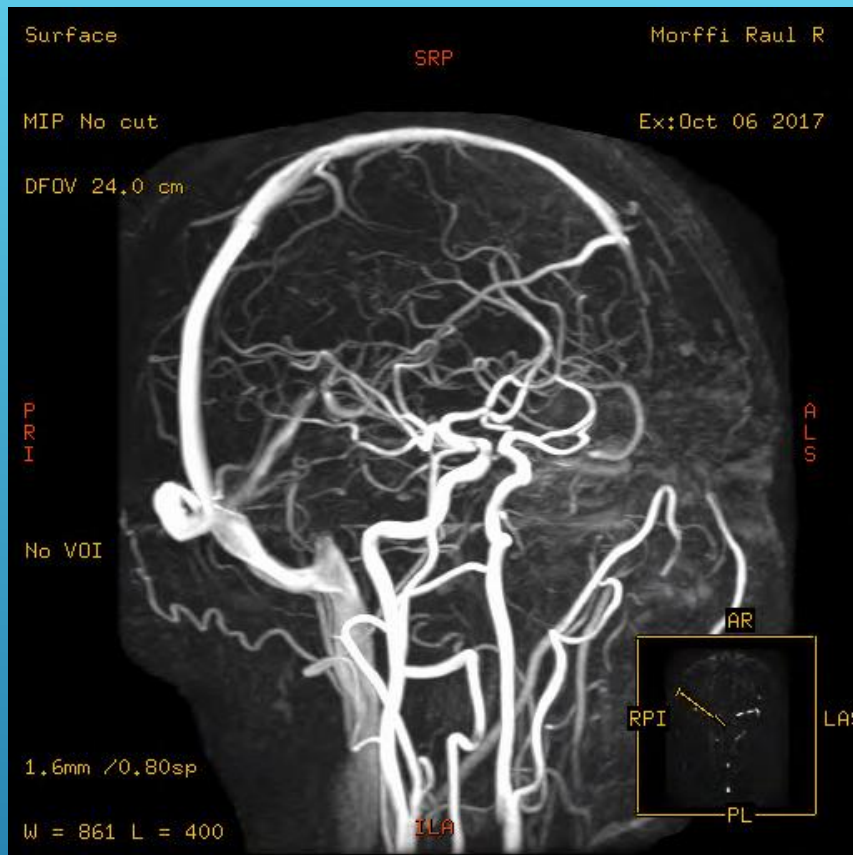
SPECIAL CONSIDERATIONS

- ▶ 66 yo healthy male presenting with worsening headache over several weeks, nausea/vomiting, but no focal deficits in 6/17
- ▶ Was being treated by infectious disease with regular IVIG for recurrent sinus infections, but not currently thought to have infection
- ▶ He was started on a heparin gtt, transitioned to warfarin and cessation of IVIG. Also received steroids for the headache, as well as Toradol, norflex and depacon
- ▶ Hypercoagulable work up is negative

CASE STUDY #1



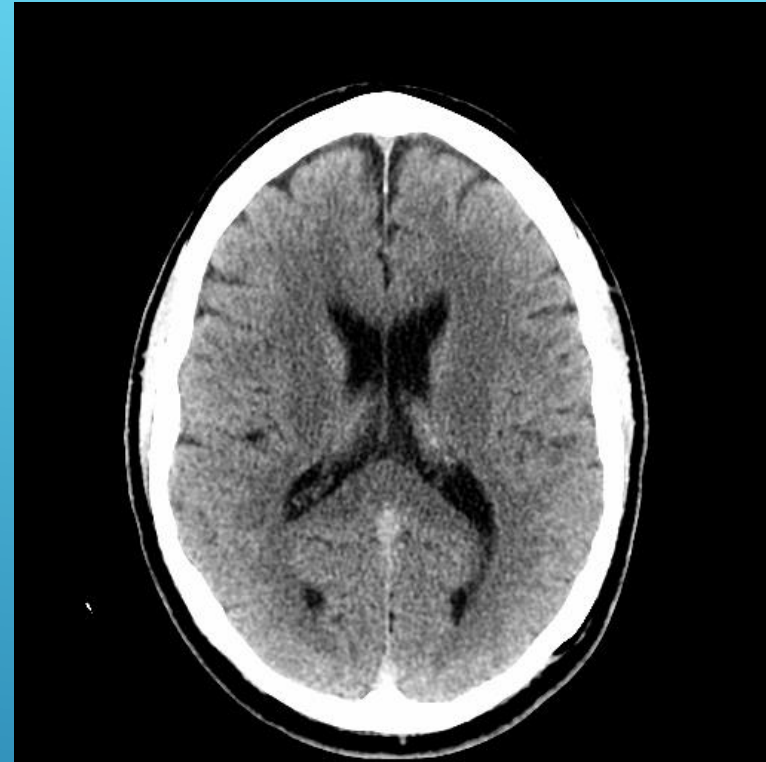
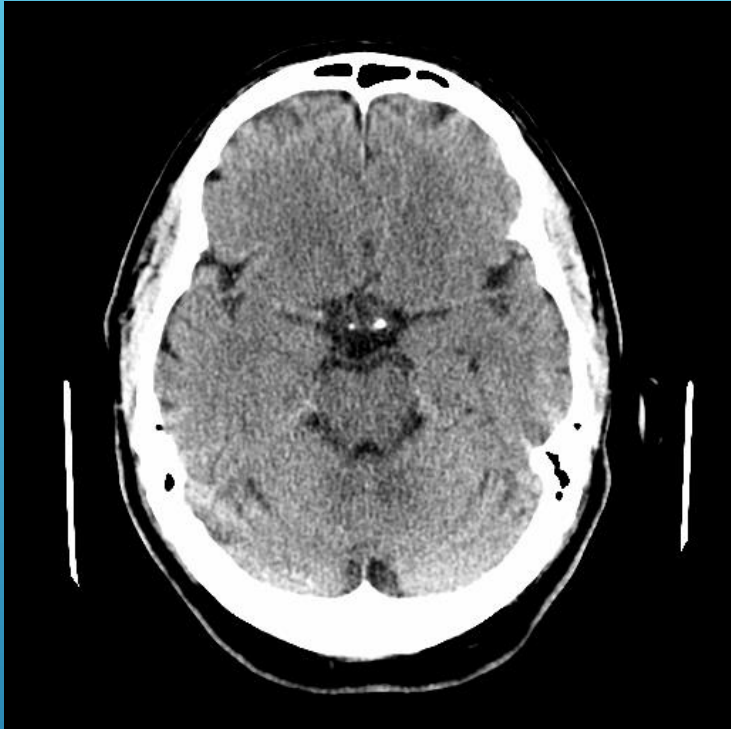
HYPERATTENUATION ALONG THE SUPERIOR SAGITTAL SINUS, RIGHT TRANSVERSE SINUS, AND STRAIGHT SINUS COMPATIBLE WITH DURAL VENOUS SINUS THROMBOSIS IN THESE SEGMENTS



PERSISTENT FOCAL RIGHT TRANSVERSE AND SIGMOID SINUS NARROWING. THE LEFT TRANSVERSE SINUS IS HYPOPLASTIC AND/OR STILL PARTIALLY OCCLUDED. FINDINGS ARE OVERALL SIMILAR TO THE PRIOR STUDY. (OCT VS DEC)

- ▶ He was treated with warfarin for approximately one year, at which time it appeared the thrombosis had resolved, but thought the patient did have hypoplastic left transverse sinus and narrowing of the posterior portion of the superior sagittal sinus and right transverse sinus
- ▶ He had recurrence of CVST in 5/21. At this time, he was not receiving IVIG
- ▶ The decision was made to continue DOAC for life—he is currently doing well on Eliquis
- ▶ We are considering a cerebral venogram to determine thrombus vs hypoplasia vs stenosis

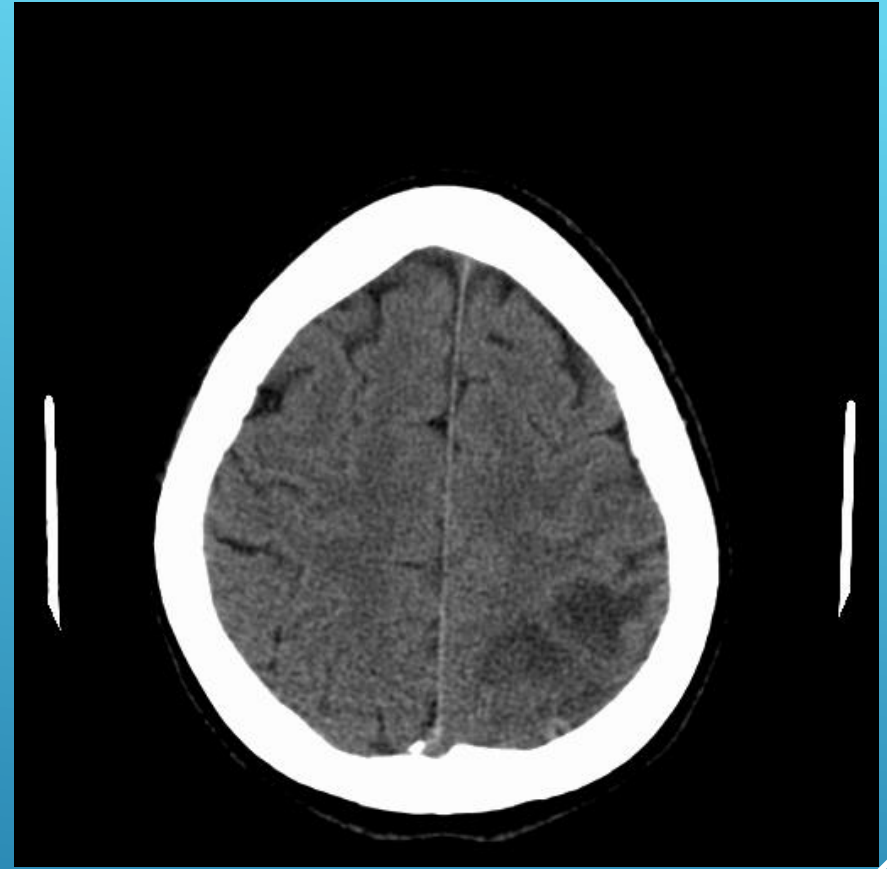
CASE STUDY #1



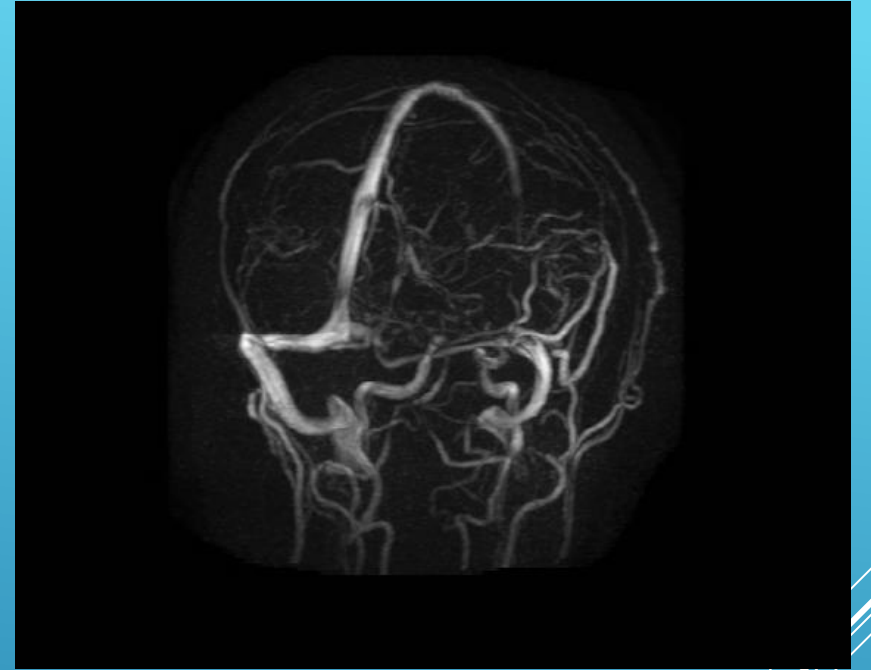
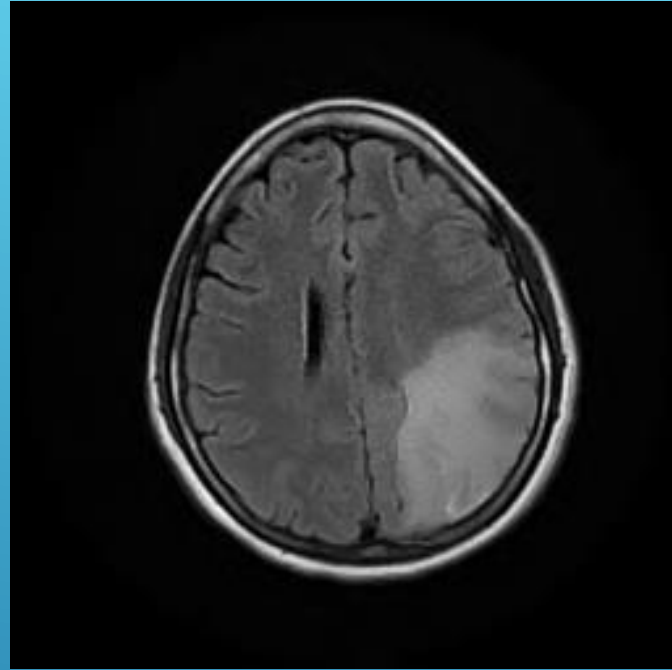
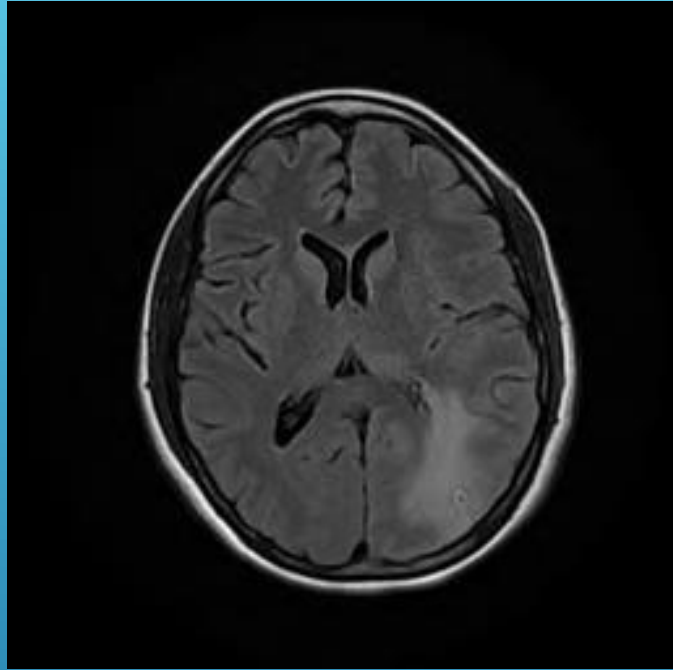
HYPERATTENUATION IN THE SUPERIOR SAGITTAL SINUS AND PROXIMAL RIGHT TRANSVERSE SINUS IS WORRISOME FOR RETHROMBOSIS OF THE DURAL VENOUS SINUSES.

- ▶ 37 yo presenting 3 days after uncomplicated vaginal delivery of a healthy infant with headache, AMS, witnessed seizures; had received an epidural with post-procedure ha which had worsened since discharge.
- ▶ No history of thrombosis, no family history of thrombosis
- ▶ Was having abnormal movements of her extremities, paresthesias on the right, rightward gaze deviation and aphasia

CASE STUDY #2

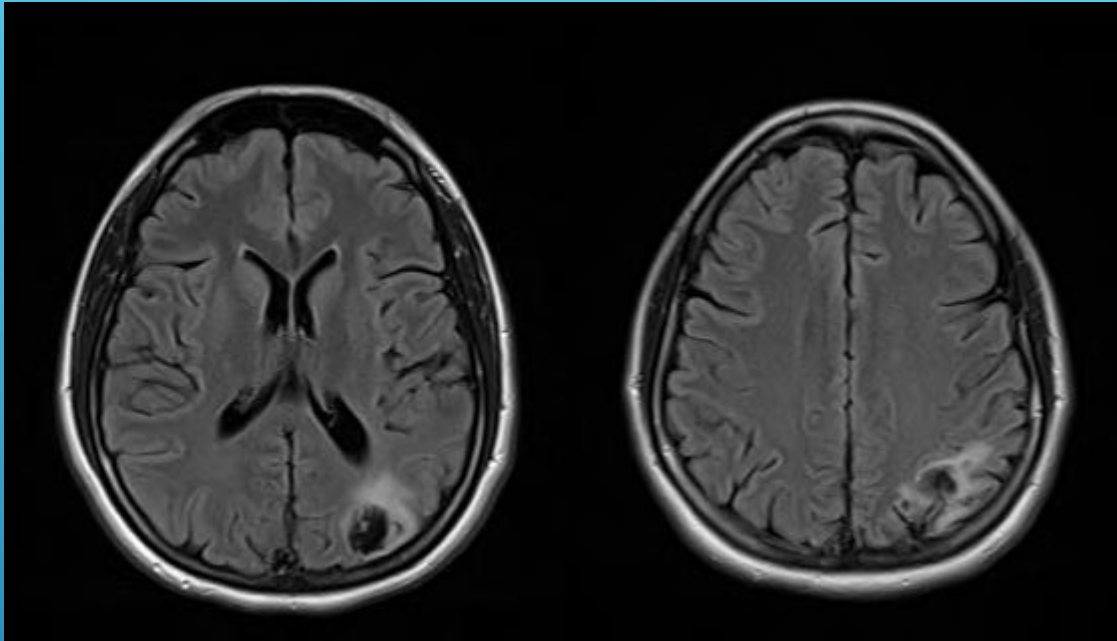


ACUTE INFARCT WITHIN THE LEFT OCCIPITOPARIETAL REGION.
RECOMMEND FURTHER WORKUP WITH MRI/MRA/MRV BRAIN. NOVEMBER
2021



DECEMBER 2021

She was readmitted with concern for continued seizures, also with psychosis, and required change in AED medication and steroids. Remained on Eliquis for anticoagulation



AUGUST 2022

Overall, she is doing well. Her baby is 10 months old, she remains on Eliquis with no issues, but is requiring 3 drug regimen to control seizures—vimpat, onfi and trileptal

- ▶ CVST often causes insidious headache followed by signs of increased ICP or focal deficits
- ▶ Seizures are common
- ▶ High index of suspicion in young women on OCP
- ▶ Ask about recent vaccinations, illnesses
- ▶ Start treatment early, heparin gtt even if ICH is present (unless VITT)
- ▶ Treat seizures
- ▶ May have both vasogenic and cytotoxic edema--? Steroids, 3% saline, etc
- ▶ Prognosis is generally good if treatment is started early

RECAP/CONCLUSION

- ▶ <https://www.hopkinsmedicine.org/health/conditions-and-diseases/cerebral-venous-sinus-thrombosis>
- ▶ <https://www.yalemedicine.org/conditions/cerebral-venous-sinus-thrombosis-cvst>
- ▶ <https://doi.org/10.1161/STROKEAHA.122.038955>
- ▶ <https://pn.bmj.com/content/20/5/356>
- ▶ <https://www.ahajournals.org/doi/10.1161/STROKEAHA.121.035564>
- ▶ <https://doi.org/10.1161/STROKEAHA.121.037541>
- ▶ https://doi.org/10.1161/str.52.suppl_1.P98
- ▶ <https://dx.doi.org/10.1136/practneurol-2019-002415>
- ▶ *Postgrad Med J* 2000;76:12-15, Holger and Abbott, "Cerebral Venous Sinus Thrombosis"
- ▶ *Google images*

REFERENCES