Diagnosis and Treatment of a Type IV Cognard Dural Arteriovenous Fistula leading to Large Volume Intracranial Bleeding in a Patient with Von Willebrand Disease

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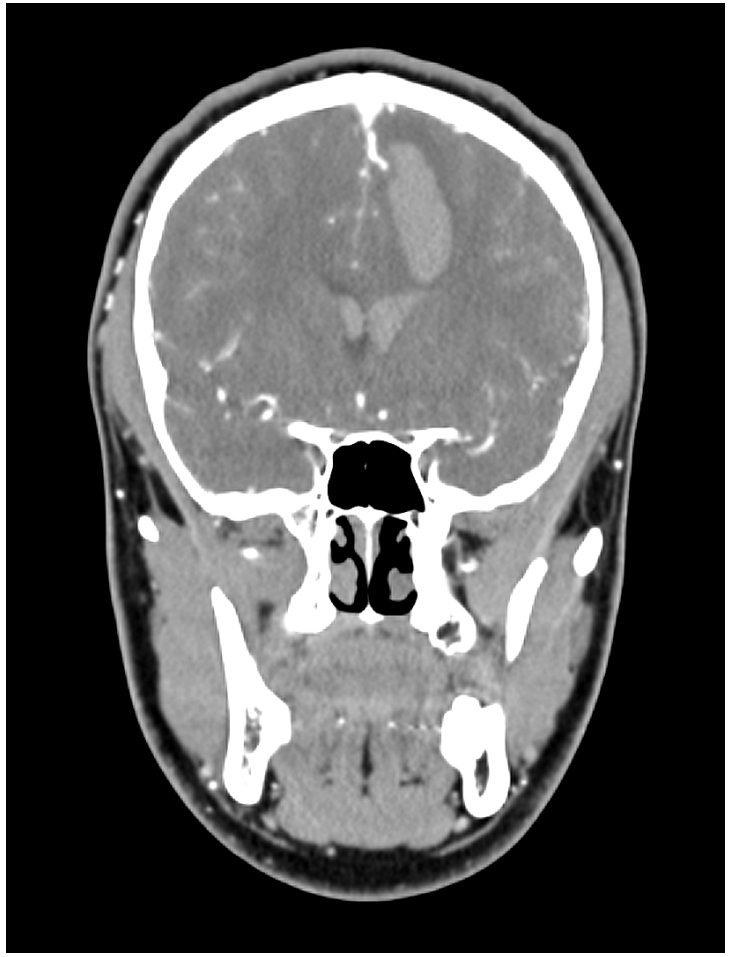
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Clinical Presentation/Initial Imaging

- 24 year old male presents with acute right hemiparesis
- Heavy alcohol and cocaine use prior night
- PMH notable for type 2 von Willebrand Disease
- Stroke alert called prompting an initial head CT
- CT Head
    - Large intraparenchymal hemorrhage within the left superior frontal gyrus with extensive intraventricular extension



Imaging Findings: CTA Head and Neck

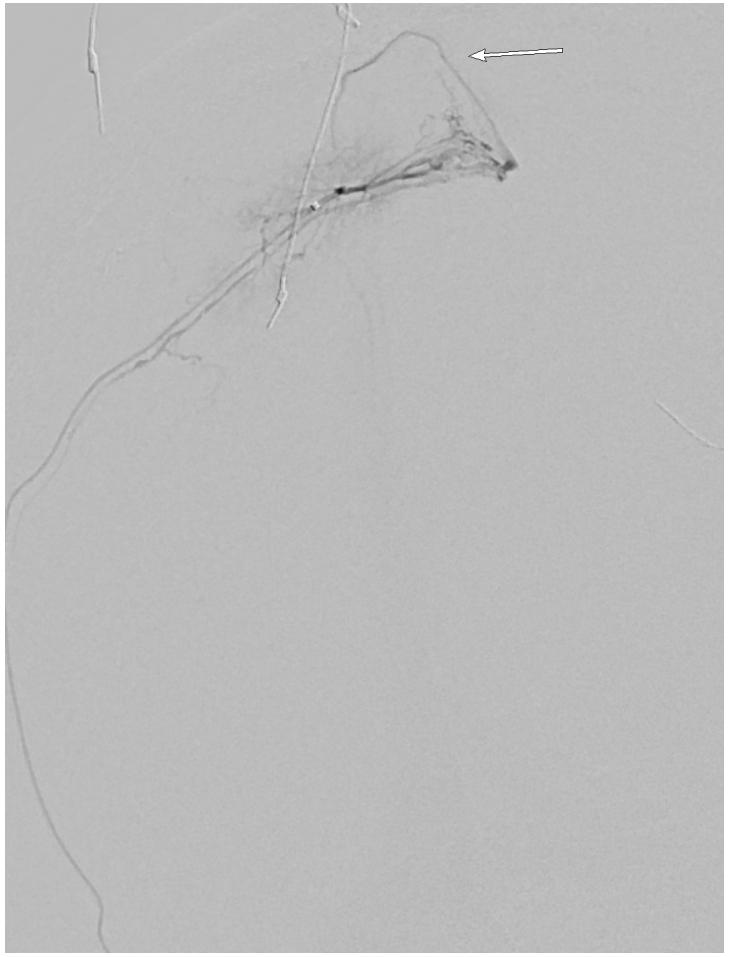




- CTA demonstrated a dilated tubular vascular structure overlying the left frontal lobe with apparent drainage towards the superior sagittal sinus, suspicious for an underlying dural arteriovenous fistula (dAVF) or other vascular malformation.
- No intracranial aneurysm was identified.

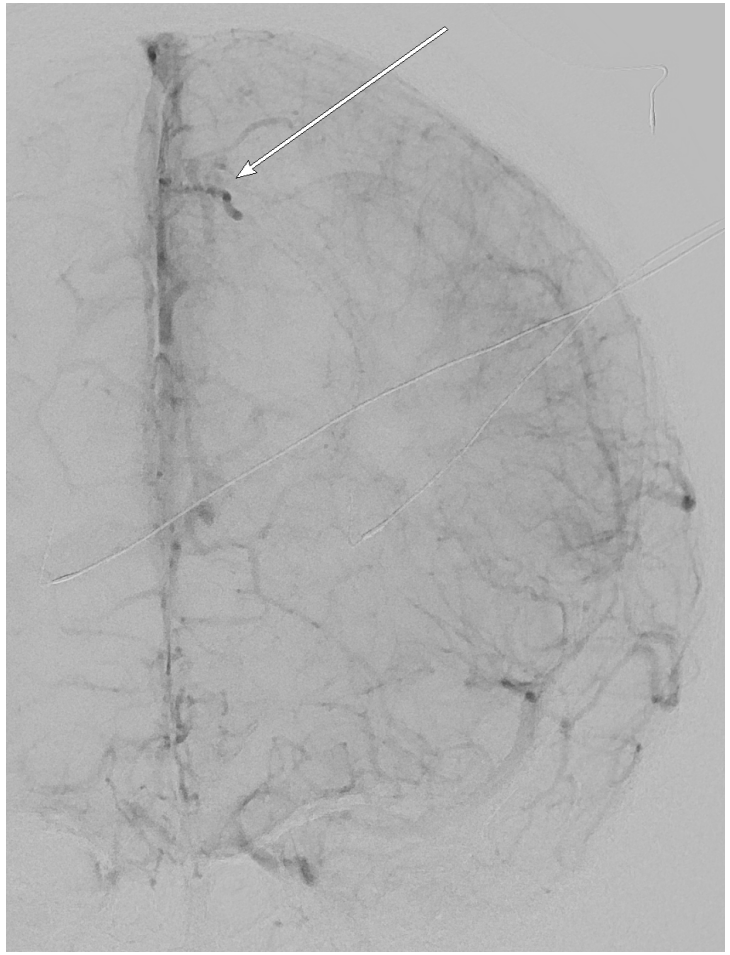
Imaging: Right ECA Diagnostic Cerebral Angiography

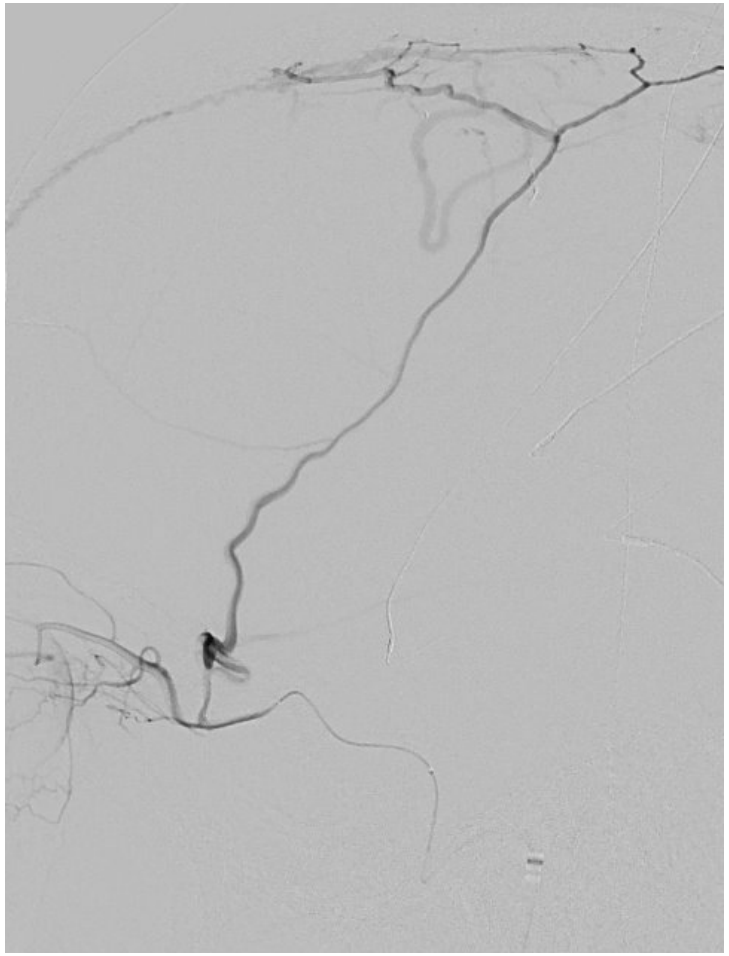




- Diagnostic cerebral angiography demonstrated a dAVF supplied by the right middle meningeal artery.
- Fistula is seen draining into an ectatic cortical vein leading into the superior sagittal sinus, consistent with a Cognard type IV dAFV

Imaging: Left Diagnostic Cerebral Angiography

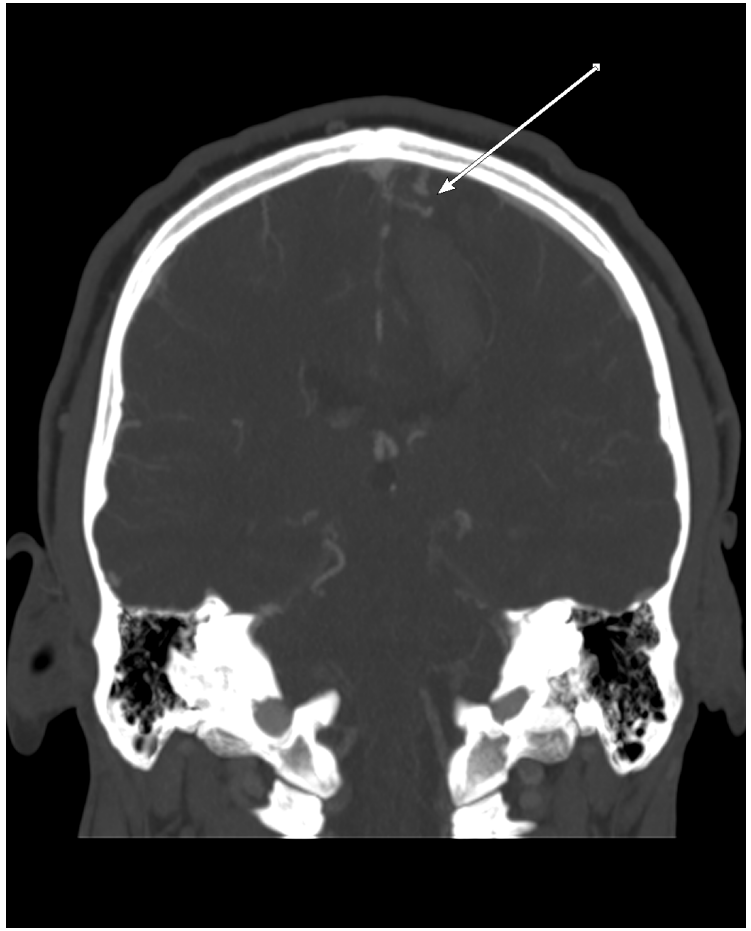




- Initial left arterial catheterization showed additional supply of fistula by a left sided branch vessel
- Catheterization of left ophthalmic artery showed an accessory left middle meningeal artery branch arising from the left ophthalmic artery, also supplying the dural AVF.

Treatment: Right middle meningeal artery branch embolization



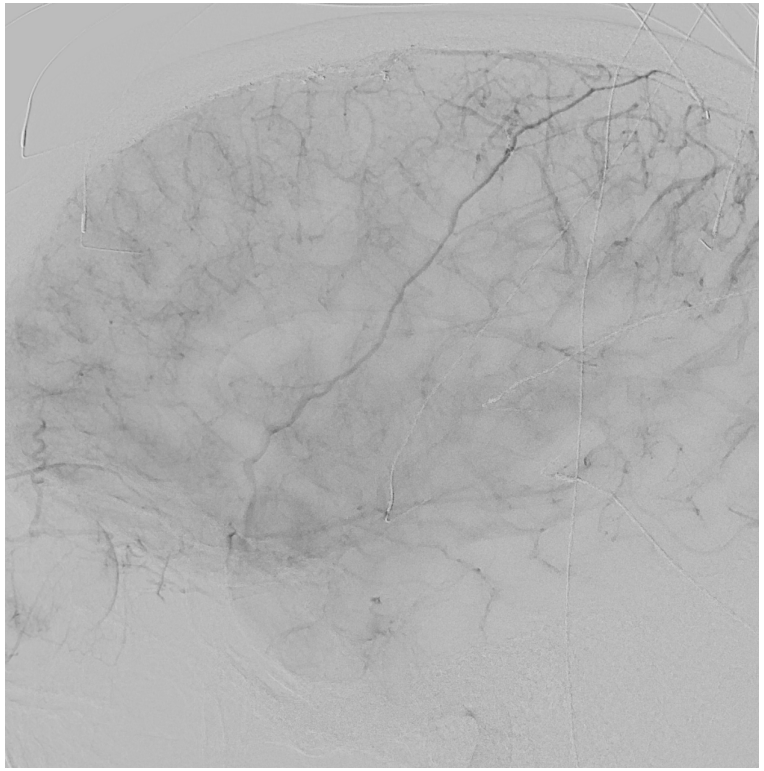


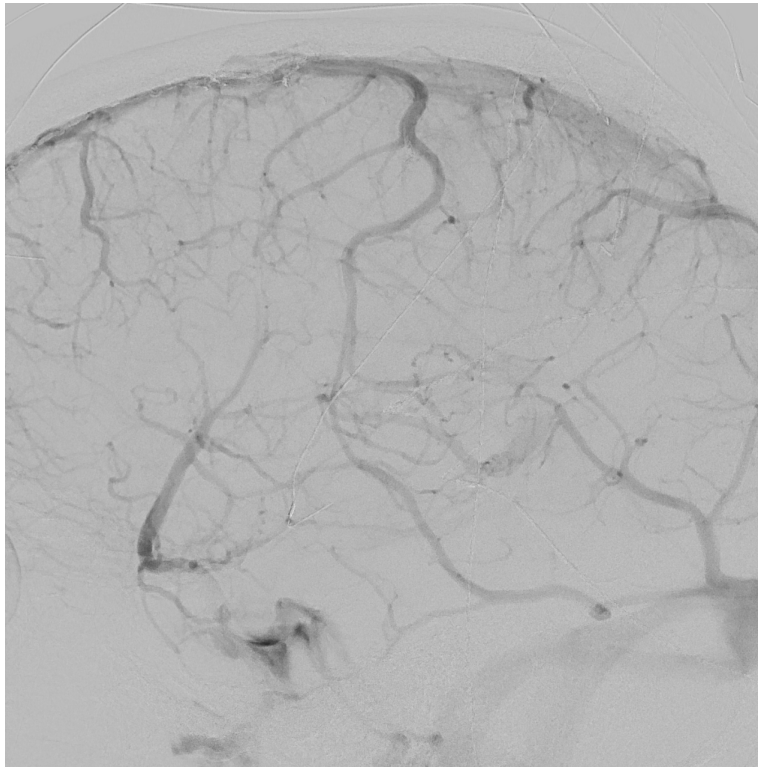
- Patient first underwent right MMA branch embolization with Onyx liquid embolic
- Post treatment cerebral angiography images show no residual right sided dAVF feeders
- Left accessory MMA feeder treatment was initially deferred due to higher risk associated with left ophthalmic branches
- Follow-up CTV showed persistent venous ectasia from presumed left sided supply
    - Prompted decision for follow-up left accessory MMA embolization

Treatment: Left accessory meningeal artery embolization

- Initial transvenous catheterization was attempted but the culprit vein was unable to be accessed (not shown)
- Left accessory meningeal artery was accessed through the left ophthalmic artery and successfully treated with Onyx liquid embolic agent
- Follow-up images including late venous phased angiography showed no residual dAVF or ectatic veins







Management

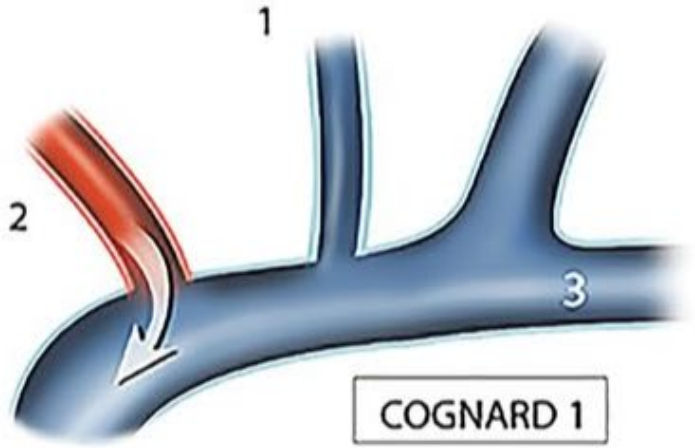
- Patient's von Willebrand disease managed with twice-weekly recombinant von Willebrand factor replacement therapy (Vonvendi)
- Monitored in ICU with serial CT heads. No new or increasing areas of hemorrhage throughout week.
- Discharged to inpatient rehabilitation and subsequently returned home

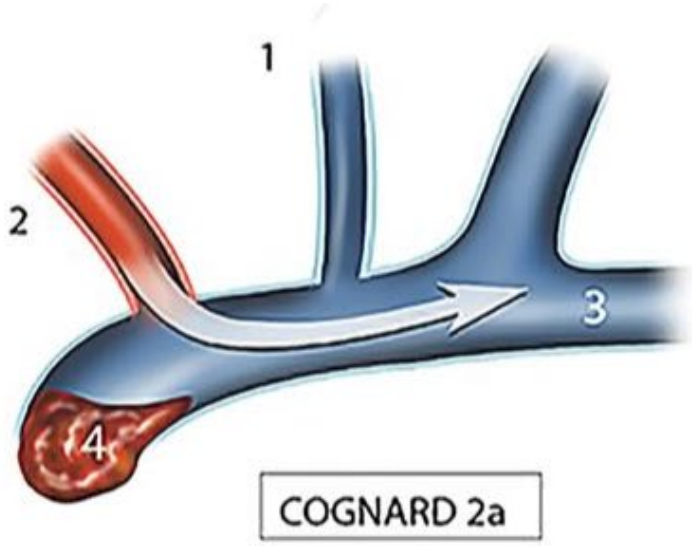
Discussion

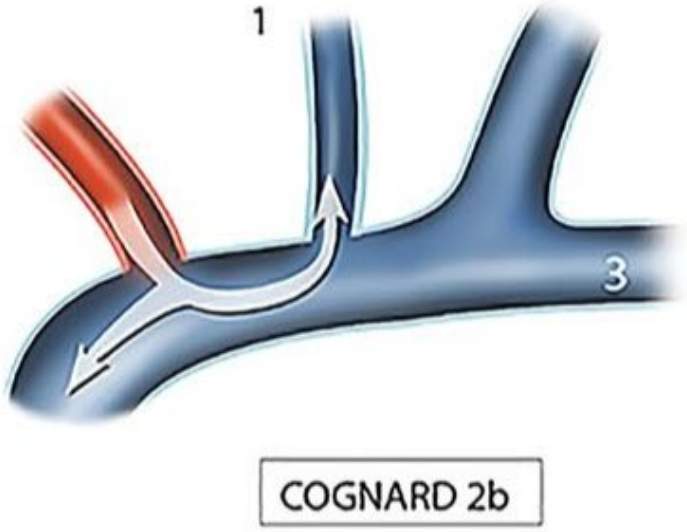
- Case highlights an unusual case of intracranial hemorrhage
- Three potentially synergistic risk factors for hemorrhage
  - Cocaine use
  - Von Willebrand Disease
  - High risk dural AVF
- Cognard Classification: Common way to classify dural AVFs and guide management decisions
  - Types 1 and 2a: lower bleeding risk so often not treated
- Others usually treated with endovascular embolization
  - Can also consider stereotactic radiosurgery or microsurgical disconnection if unable to access endovascularly

Cognard Classification: Types 1 and 2

1. Cortical vein
2. Artery
3. Sinus
4. Thrombus

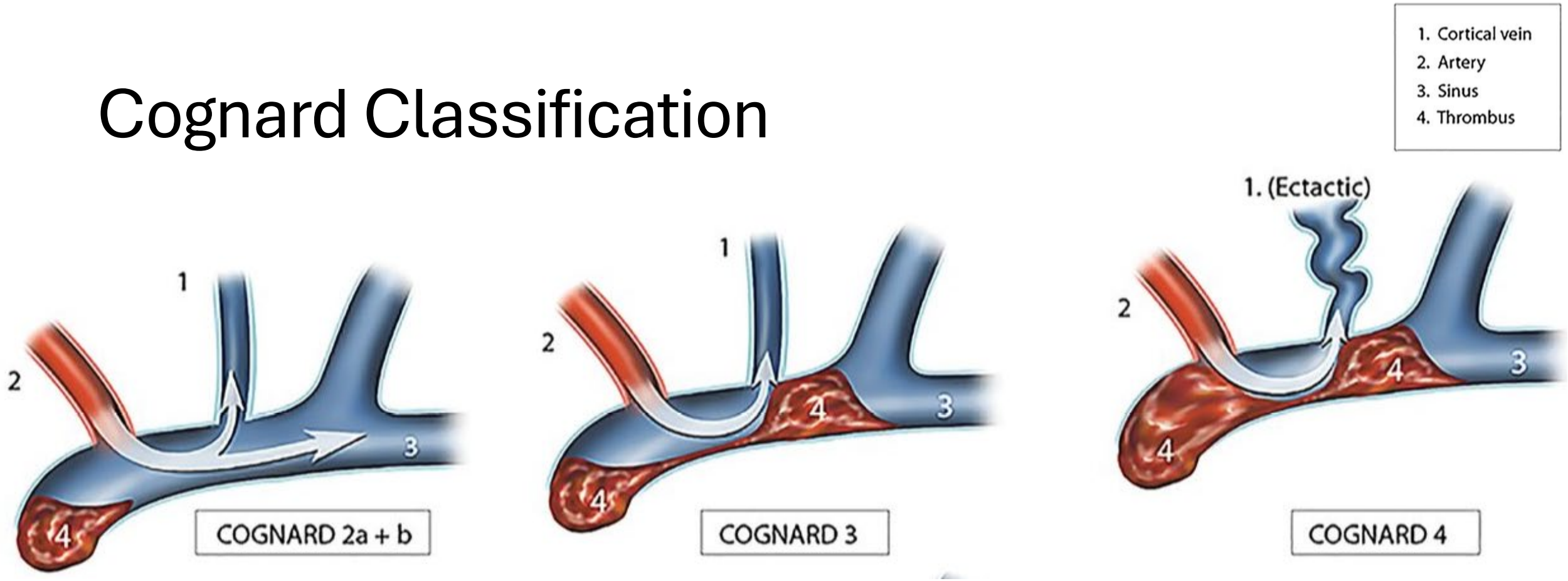






- **Type 1**
  - Antegrade flow through sinus
  - No Reflux into cortical vein
- **Type 2a**
  - Retrograde flow through sinus
  - No Reflux into cortical vein
- **Type 2b**
  - Antegrade flow through sinus
  - Reflux into cortical vein

Cognard Classification



- **Type 2a + b**
  - Retrograde flow through sinus
  - Reflux into cortical vein
- **Type 3**
  - Drainage directly into cortical vein
  - No significant drainage through a sinus
- **Type 4**
  - Drainage directly into an ectatic cortical vein

Cognard Classification

- Type 5
  - Arterial drainage into a spinal perimedullary vein
  - Bypasses normal cerebral veinous drainage
  - Can cause Venous congestion and edema within spinal cord
  - Often leads to myelopathy

