ARUBA Study: Preliminary Variable Definitions

Demographic Variables

The patient's birth date, residential ZIP-code, gender (male/ female), self-defined race/ethnicity (American Indian, Asian, black, Hispanic, white, other), current insurance status (Medicaid, Medicare, Private Insurance, any combination, other) and enrollment date are documented. Age at presentation is calculated as the patient's age (in years) at the time of the initial AVM presentation (diagnostic event).

Clinical Variables

The diagnostic event (or initial AVM presentation) is defined as the clinical picture of the index event that brought the patient to a medical encounter which directly led to the discovery of the AVM. The date of the diagnostic event is documented and the mode of clinical presentation further stratified into:

Intracranial Hemorrhage refers to bleeding into the brain or its surrounding spaces. A hemorrhagic presentation (or incident intracranial hemorrhage) is defined as a clinically symptomatic (i.e. stroke-like) event with signs of fresh or recent intracranial blood on head CT and/or MRI or in the cerebrospinal fluid. The primary bleeding location is further classified as being parenchymatous, subarachnoid, intraventricular, or any combination.

Any event of clinical seizure activity is syndromatically stratified into simple focal, partial complex, and (primary or secondary) generalized seizures.

A focal neurological deficit refers to a functional deficit on clinical exam and is stratified as to whether the deficit was persistent, progressive or reversible at the time of evaluation.

Headaches are further stratified into (1) sudden onset headache, (2) migraineous headaches (typical features required, i.e. with or without aura, typical course, vegetative symptoms, etc.), (3) non-specific remittent headaches, and (4) chronic headaches (≥4days/week).

Other modes of presentation include any other AVM-related symptoms (e.g. a bruit) which eventually lead to the diagnosis of the malformation.

Incidental or asymptomatic presentation refers to a clinical presentation that was clearly unrelated to the AVM regarding the indication for imaging, e.g., pituitary gland dysfunction or chronic sinusitis.

Handedness (right-handed, left-handed, ambidextrous) will be coded according to the patient's self-definition in the neurological history.

Measures of health-related dysfunction include Rankin Scale, Barthel index, and EuroQol and will be documented for each patient at each evaluation.

Morphological Variables

The imaging source and date of the computed tomography, magnetic resonance imaging, arterial magnetic resonance angiography , diagnostic 4-vessel brain angiography and/or superselective cerebral angiography, will be documented. The nearest imaging source in time to the patient's presentation are the basis for the description of morphological variables.

Brain AVM side (right, left, midline) refers to the topographic location in cases where one malformation has been detected. For those harboring ≥1 brain AVM, each malformation will be coded separately.

The AVM size (nidus size) is measured as the largest diameter in millimeters based on pretreatment MRI and/or cerebral angiogram.

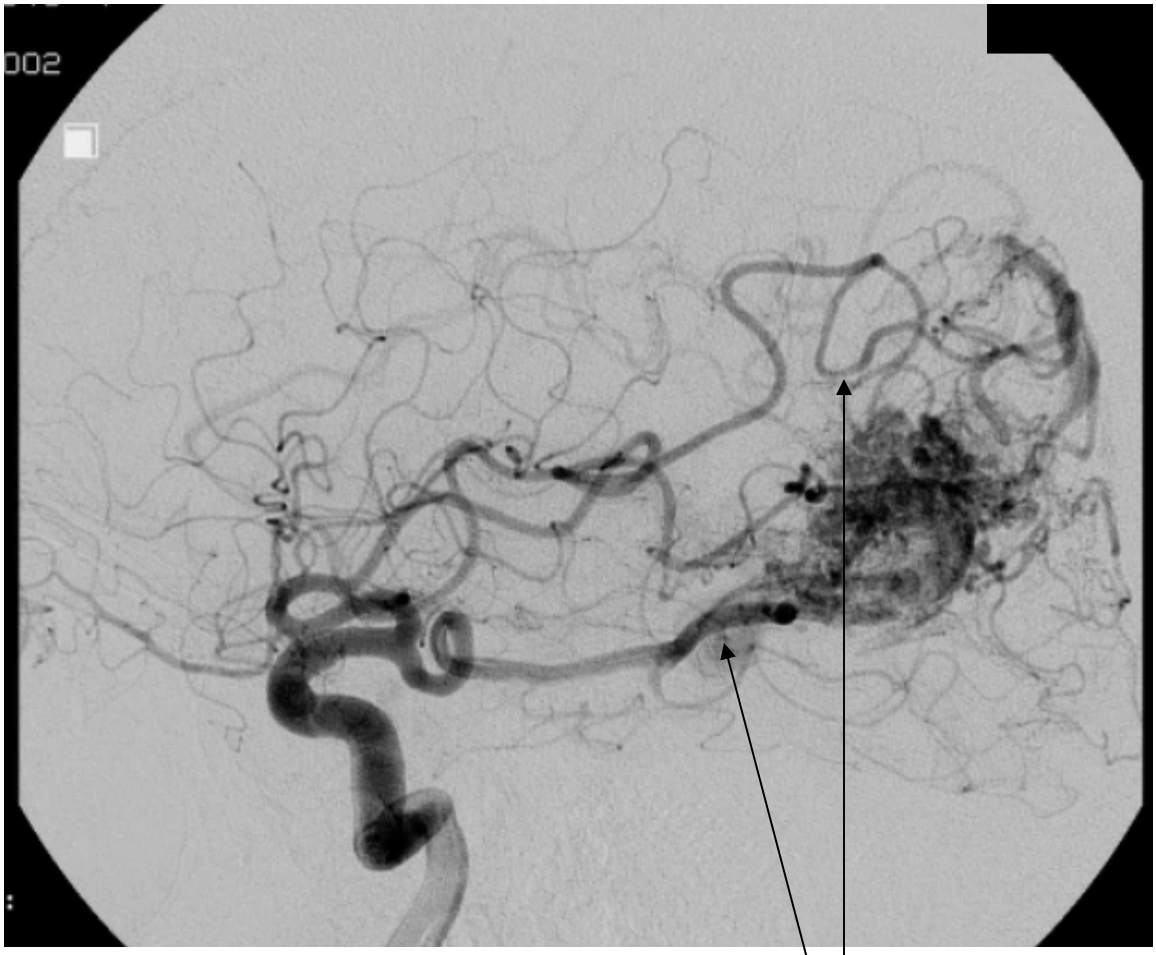
Anatomic AVM location is stratified into lobar (i.e. frontal, parietal, temporal, and/or occipital lobe), deep (basal ganglia, thalamus, internal capsule, corpus callosum) and/or infratentorial (midbrain, pons, medulla, cerebellum) location.

A so-called "eloquent" brain location (as defined by the Spetzler-Martin scale) is coded positive in cases where the AVM is located in "the sensorimotor, language, and visual cortex; the hypothalamus and thalamus; the internal capsule; the brainstem; the cerebellar peduncles; and the deep cerebellar nuclei". [1]

An AVM feeding artery is defined as any intracranial vessel that angiographically contributes arterial flow to the malformation. Feeding arteries may be "parent' arteries, i.e. that give rise to vessels that directly or indirectly supply flow to the AVM. Coding of multiple vessels is possible. For documentation, single feeding arteries will be stratified into right or left ICA, AChA, ACA cortical branches, ACA penetrators, MCA cortical branches, MCA penetrators, VA, PICA, BA, AICA, SCA, PCA cortical branches, PCA perforators, PChA, any dural supply (via ECA or VA branches).

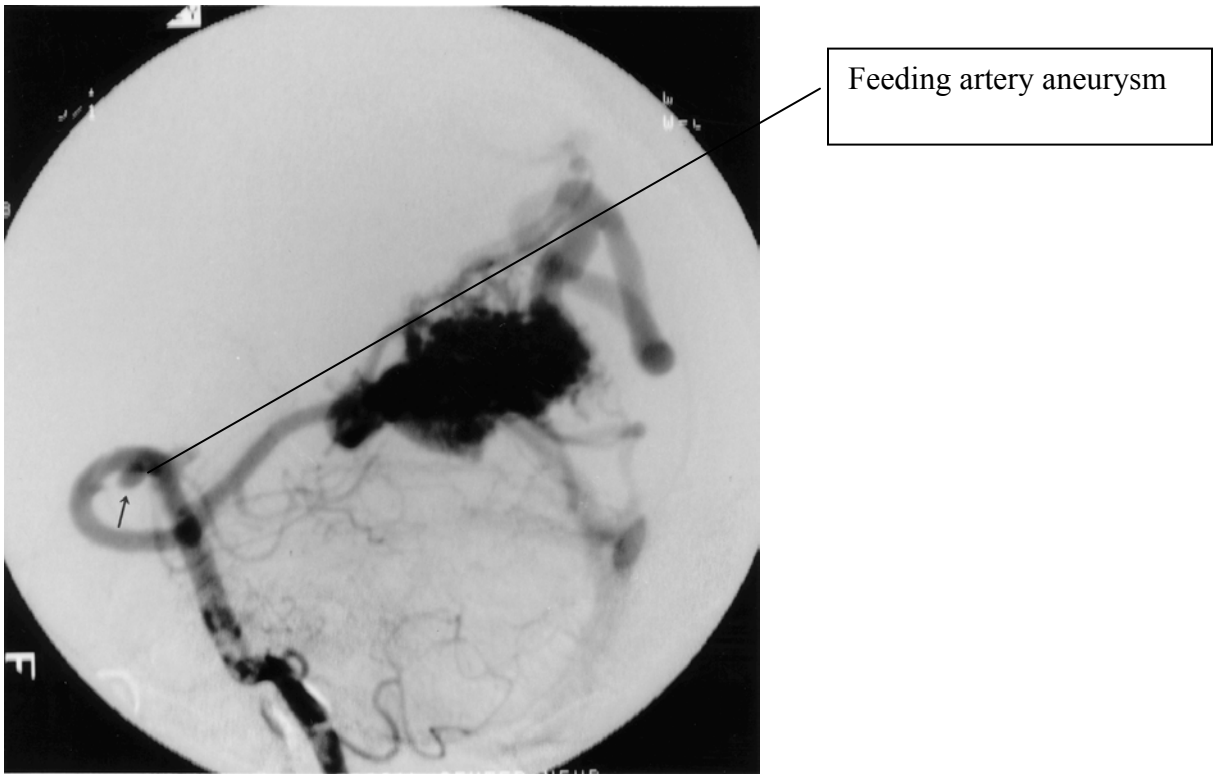
Moyamoya type changes are defined as any pattern of collateral small vessel recruitment due to proximal feeding artery stenosis or occlusion.

Borderzone location applies to AVMs located in the inter-arterial region shared by the distal anterior, middle, and/or posterior cerebral arteries. Borderzone AVMs are commonly supplied by branches of at least 2 of the individual major circle of Willis arteries, i.e. the anterior and middle; middle and posterior; anterior and posterior; or anterior, middle, and posterior cerebral arteries.

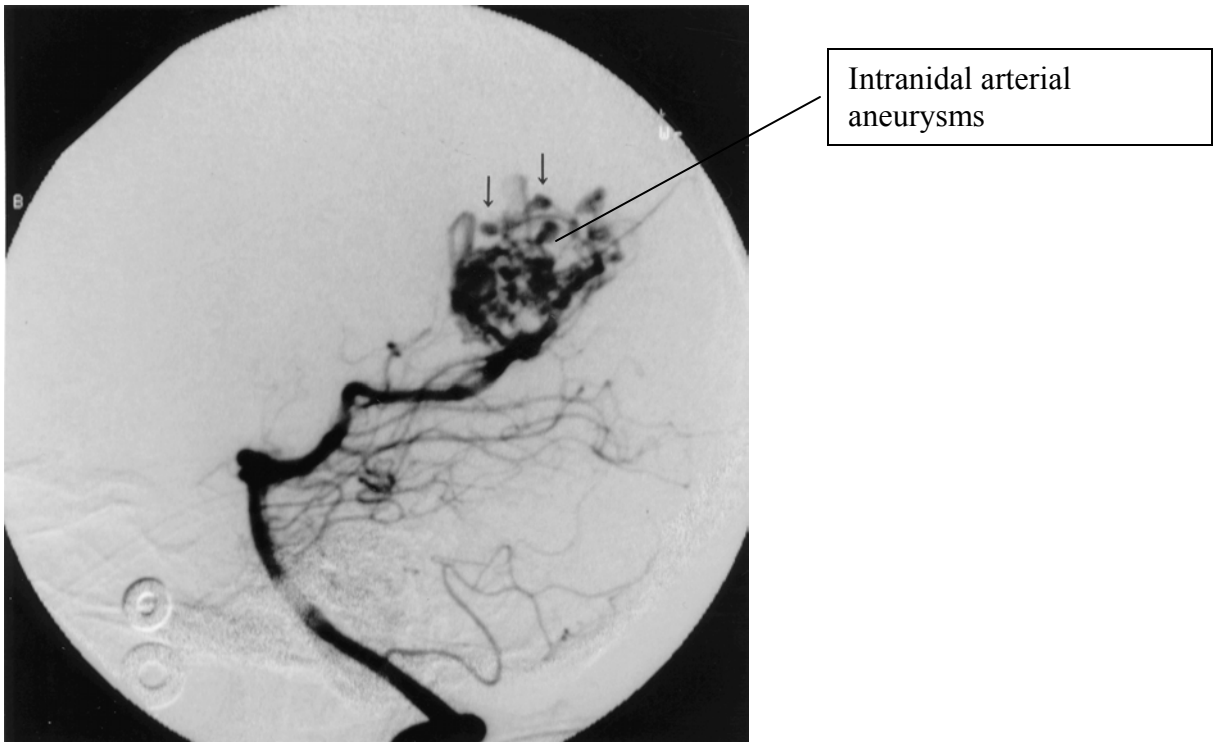


AVM located in the arterial Borderzone between MCA and PCA branches (PCA flow via Pcom)

Concurrent arterial aneurysms are defined as saccular dilatations of the lumen ≥2 times the width of the arterial vessel that carried the dilatation and further classified as (1) feeding artery aneurysms, (2) intranidal aneurysms, and (3) aneurysms unrelated to blood flow to the AVM. The number and vessel location of any aneurysm subtype will be documented.

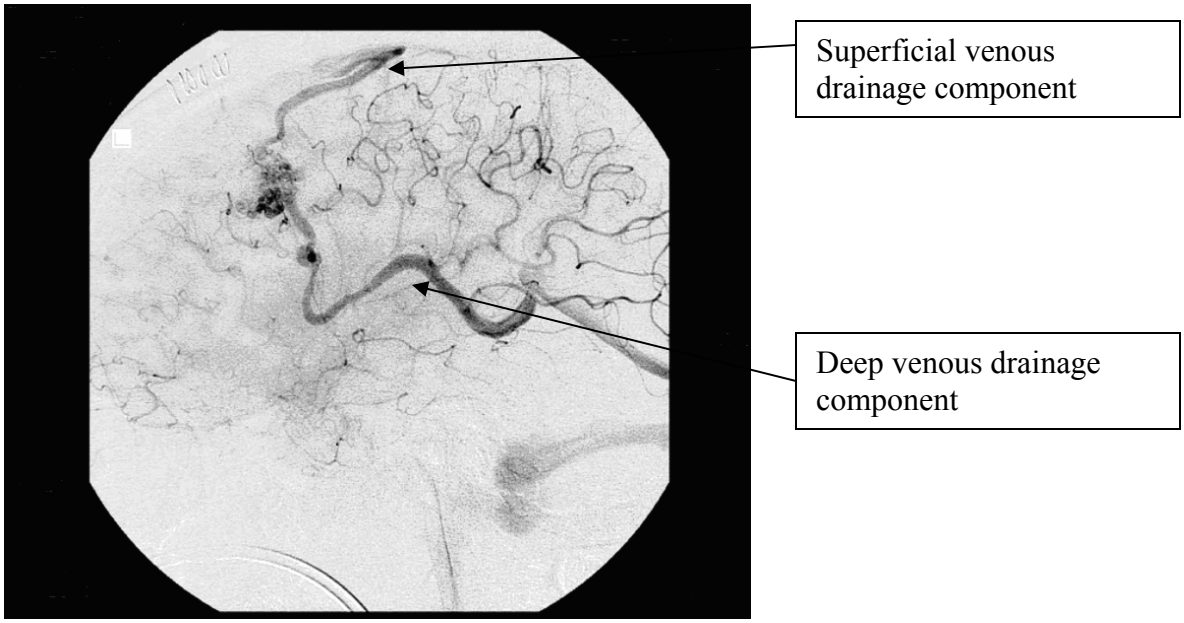


Intranidal aneurysms are coded only when visualized early after angiographic injection, e.g. before substantial venous filling had occurred.



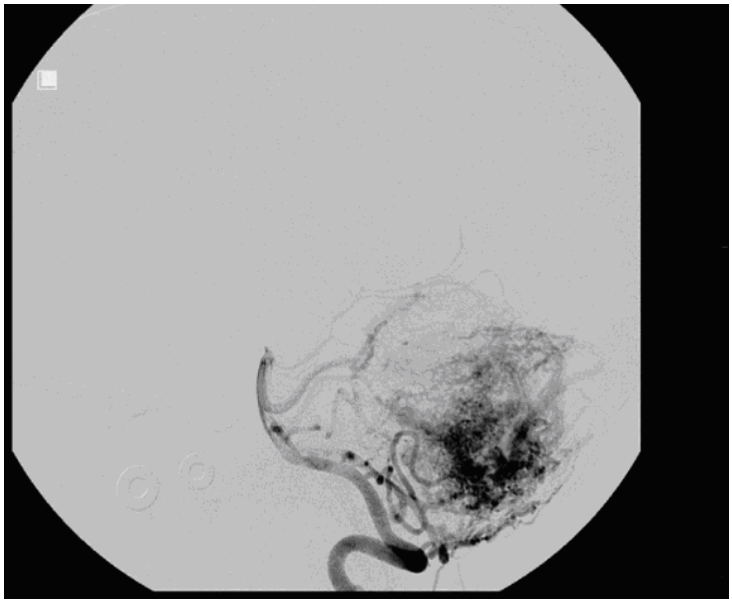
Infundibula, arterial ectasias (i.e. dilated feeding vessels), and intranidal aneurysmal dilatations seen during the venous angiographic phase only will not be coded as arterial aneurysms. Arterial aneurysms will be coded as unrelated to the AVM when located on intracranial arteries not contributing blood flow to the AVM.

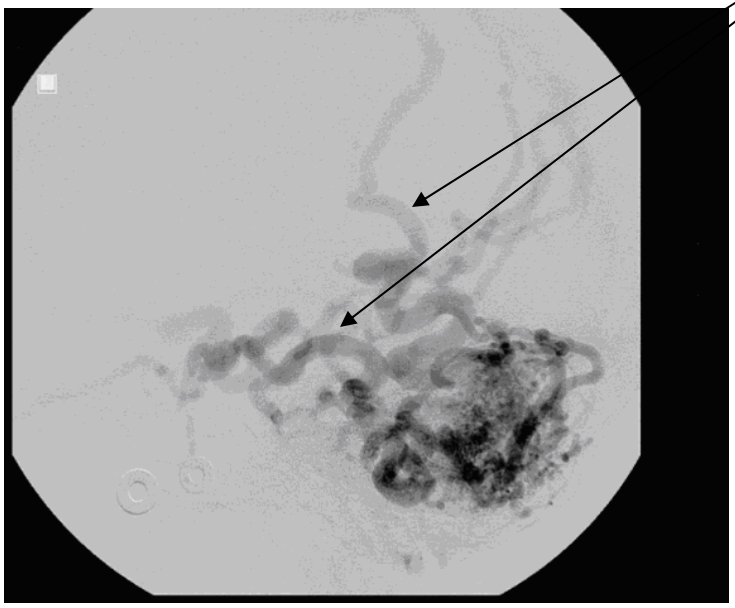
The venous drainage pattern is categorized as angiographic drainage into the superficial cortical veins (superficial venous drainage), drainage into the deep venous system (deep venous drainage such as the internal cerebral veins, basal veins, vein of Galen, etc.), and combined superficial and deep drainage.



"In the posterior fossa, only cerebellar hemispheric veins that drain directly into the straight sinus, torcula, or transverse sinus are considered to be superficial."[36]

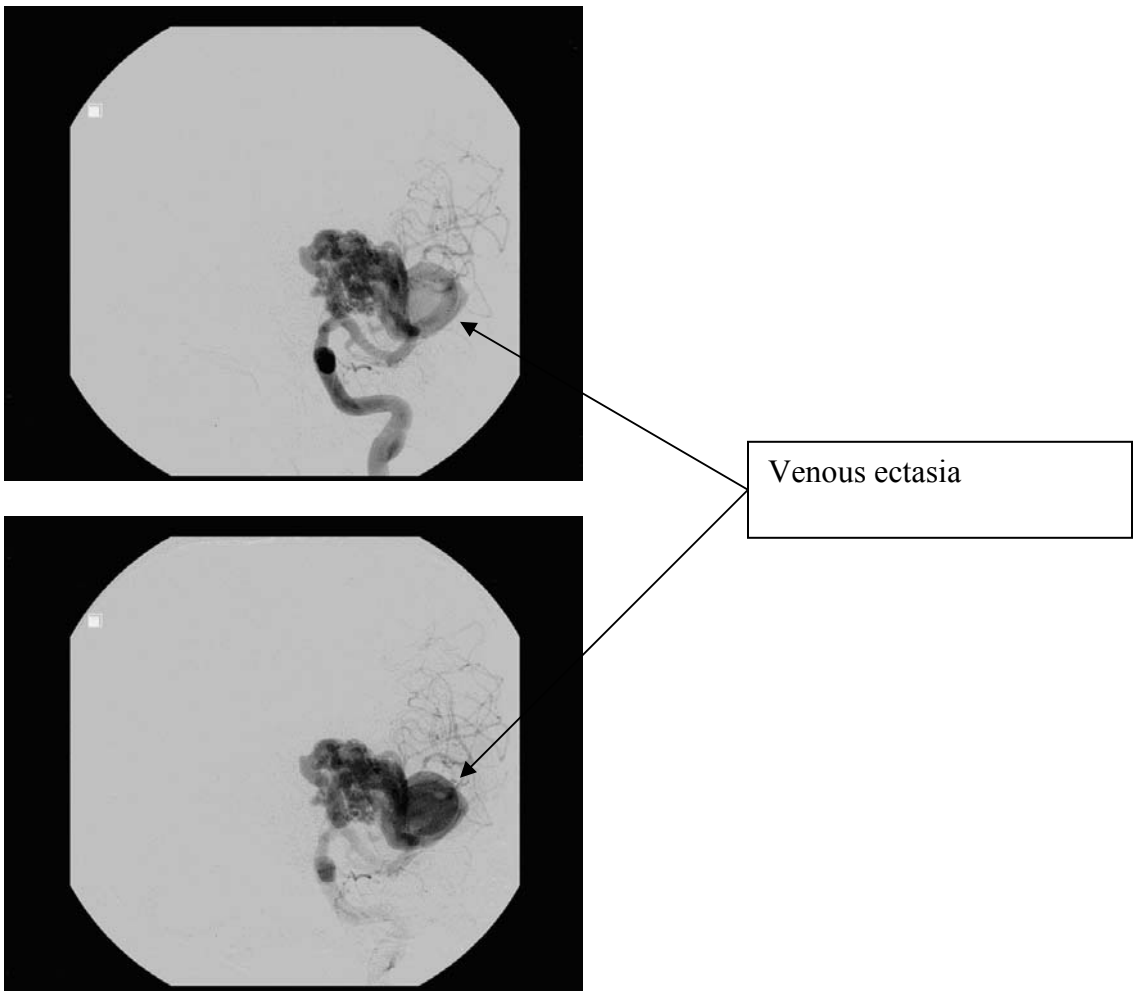
Venous stenosis / occlusion is defined as a ≥2-fold caliber narrowing or occlusion of any draining vein outflow pathway seen in two angiographic views.





Venous outflow obstruction with various venous collaterals

Venous ectasia is coded positive in cases with a ≥2-fold caliber increase change in any draining venous channel.



Treatment variables

The date of each treatment procedure is coded along with the treatment modality (i.e. surgery, endovascular treatment, or radiation therapy) and treatment target (AVM treatment, aneurysm treatment). Each procedure is labeled as being technically complete or incomplete.

Clinical variables and measures of health-related dysfunction (as outlined above) are reassessed after each procedure.

Reference:

1. Spetzler RF. Martin NA. A proposed grading system for arteriovenous malformations. J Neurosurg 1986;65:476-483