Cranial Vessel Embryology and Imaging Anatomy

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This publication is endorsed by: European Society of Neuroradiology (www.esnr.org).

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F. Barkhof et al. (eds.), *Clinical Neuroradiology*,
https://doi.org/10.1007/978-3-319-68536-6_20

Abstract

The brain vascularization is extremely complex with a large range of possible vascular variants, providing for each individual an exclusive arterial and venous system configuration.

The overview of the arterial and venous brain vascular development presented is essential for the understanding of some vascular dispositions, such as the persistence of embryonic vessels/vascular patterns and complex vascular malformations, like the metameric syndromes.

The review of the fundamental aspects of the arterial circulation, including the description of the anterior and posterior circulation, arterial variants, and territories, provides the basis for the understanding of the diseases later discussed in other chapters.

The intracranial venous system is also overviewed, encompassing the dural sinus anatomy, the supratentorial and infratentorial superficial and deep venous anatomy, and the venous anastomosis. The development venous anomalies and sinus pericrannii are also discussed.

Radiology has contributed tremendously for the recent advances in brain vascular anatomy knowledge using the acquisition of high-resolution 3D DSA images and supraselective catheterization during endovascular procedures. Clinical neuroradiology plays a major role for the evaluation of brain vascular anatomy that is essential for the diagnosis and treatment (surgical or endovascular) of vascular diseases.

Keywords

Vasculogenesis · Angiogenesis · Arteriogenesis · Arterial development · Venous development · Vascular · Anatomy · Internal carotid artery(ies) · Vertebral artery(ies) · Basilar artery · Anterior cerebral artery(ies) · Posterior cerebral artery(ies) · Middle cerebral artery(ies) · Anterior-inferior cerebellar artery (ies) · Posterior-inferior cerebellar artery(ies) · Superior cerebellar artery(ies) · Cerebellar veins · Cortical veins · Vein of Galen · Venous sinus(es)

Abbreviations

AA	Aortic arch
ACA	Anterior cerebral artery(ies)
AChA	Anterior choroidal artery(ies)
ACommA	Anterior communicating artery
AICA	Anterior-inferior Cerebellar artery (ies)
Ang-1	Angiopoietin-1
AV	Arterio-venous
AVM	Arterio-venous malformation
BA	Basilar artery
bFGF	Basic fibroblast growth factor
CAMS	Cerebrofacial arteriovenous metameric syndromes
CMS	Cerebrofacial metameric syndromes
CoW	Circle of Willis
CT	Computed tomography
CTV	computed tomography venogram
CVMS	Cerebrofacial venous metameric syndromes
DSA	Digital subtraction angiogram
DVA	Development venous anomalies
ECA	External carotid artery(ies)
ICA	Internal carotid Artery(ies)
LPChA	Lateral posterior choroidal artery (ies)
MCA	Middle cerebral artery(ies)
MIP	Maximum intensity projection
MPChA	Medial posterior choroidal artery (ies)
MRV	Magnetic resonance venogram
PCA	Posterior cerebral artery(ies)
PChA	Posterior choroidal artery(ies)
PCommA	Posterior communicating artery(ies)
PDGF	Platelet derived growth factor

PHACES	Posterior cranial fossa malformations, hemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects, and abnormalities of the Eye
PICA	Posterior-inferior cerebellar artery (ies)
SAMS	Spinal arteriovenous metameric syndromes
SCA	Superior cerebellar artery(ies)
SMCs	Smooth muscle cells
SP	Sinus pericranii
SWI	Susceptibility weighted image
TA	Trigeminal artery(ies)
TOF	Time of flight
VEGF	Vascular endothelial growth factor

Concepts in Vessel Development: Vasculogenesis, Angiogenesis, Arteriogenesis, and Intracranial Arterial and Venous Development

Vasculogenesis, Angiogenesis, Arteriogenesis

Vascular development is a complex and dynamic process starting at a very early phase of the embryo development, shortly after gastrulation (early embryonic development to a double-wall stage). Several biological processes intervene, such as precursor cell migration to the target organs, de novo formation of primitive vessel channels (vasculogenesis), shaping of these primitive new vessels channels (angiogenesis). An intricate signaling pathway, mainly guided by vascular growth factors, such as vascular endothelial growth factor (VEGF) for vasculogenesis, basic fibroblast growth factor (bFGF) and VEGF for angiogenesis, and platelet derived growth factor (PDGF) plus angiopoietin-1 (Ang-1) for vessel stabilization. These factors are produced in response to genetic, systemic, and local (target organ) factors and are responsible for regulation of the vessel development. Local hypoxia is an important stimulus for the release of these angiogenic factors during these phases (Milner 2014; Ribatti 2015).

Blood vessels walls are composed of two main cell types, the endothelial cells and mural cells. Endothelial cells are the chief cell type for development and remodeling of the vascular system. Multiple layers of smooth muscle cells and an outer layer of pericytes compose the vessel wall of arteries and veins. Pericytes have two major functions: structural, supporting the blood vessels, and functional, forming with the endothelial cells the blood –brain barrier (Milner 2014; Ribatti 2015).

These cells have different embryological origins. The blood vessel endothelial cells originate from the mesoderm and the vessel media components from the neural crest. The mesenchymal stem cells (mesoderm) differentiate into angioblasts and hemangioblasts, which are precursors of endothelial cells and hematopoietic cells, respectively. The neural crest also gives rise to the skin, connective tissue, skeleton of the craniofacial region, and meninges (Milner 2014; Ribatti 2015).

Endothelial cells can initiate the vascular development process but are not sufficient to complete this process alone. The mural cells, especially the pericytes, the extracellular vessel wall matrix, and elastic membranes, are important for the vessel stabilization. Pericytes contribute for vessel stabilization by inhibiting endothelial proliferation and migration, and by stimulating production of extracellular matrix (Milner 2014; Ribatti 2015).

Blood vessel development includes the formation of new vessels from cellular precursors (vasculogenesis), and the growth and remodeling of the primitive vessel network into a complex vascular network (angiogenesis). The angioblasts migrate into the developing target organs, proliferate, and differentiate into endothelial cells, forming the initial blood vessels (vasculogenesis). Subsequently, the other vascular cells (the mural cells), such as pericytes, fibroblasts, mesenchymal cells, and smooth muscle cells, are recruited in response to growth factors released by the endothelial cells (Milner 2014; Ribatti 2015).

Vasculogenesis occurs at an initial stage of vascular development and is responsible for the emergence of the blood vascular system. This biological process is responsible for the blood vessels de novo formation from migrating

angioblasts. The angioblasts will proliferate and differentiate into endothelial cells, forming cords of cells and a primitive network of vascular channels (Milner 2014; Ribatti 2015).

Vasculogenesis will produce an immature and poorly functional vasculature. After vasculogenesis, several vascular maturation mechanisms occur, such as vascular branching, pruning, remodeling occur continuously, in response to local organ development, producing mature and stable vessels (Milner 2014; Ribatti 2015).

Angiogenesis always follows vasculogenesis, being the biological process of growth and reshaping of the primitive network of vascular channels formed during vasculogenesis. In arteriogenesis, there is collateral artery growth through the outgrowth of pre-existent collateral arterioles, requiring proliferation of endothelial cells and smooth muscle cells (SMCs), in their diameter to compensate for changes/disturbances in blood flow. It may occur postnatal in physiological processes like wound healing and female reproductive cycle, or in a variety of pathological processes, such as diabetic retinopathy or tumor (neo) angiogenesis (Milner 2014; Ribatti 2015).

Two different angiogenesis processes are generally described: sprouting (new vessels formed by the ramification and growth of pre-existing) and intussusception or non-sprouting (splitting of one pre-existing vessel into other vessels) angiogenesis. Sprouting angiogenesis is essential for brain vascular network, like in other ectodermal organs (Milner 2014; Ribatti 2015).

In sprouting angiogenesis, group of migratory cells, located at the tip of the primitive vessel, extends with the formation ("sprouting") of new vessels. This vessel expansion may also accommodate the incorporation of newer migrating angioblasts. Non-sprouting angiogenesis mechanisms, such as intussuscepted microvascular growth, in which there is the insertion of interstitial cell columns into pre-existing vessels, create vascular partitions (Milner 2014; Ribatti 2015).

It seems that shear stress is the main force to initiate arteriogenesis, in opposition to angiogenesis in which hypoxia is a leading stimulus. Hemodynamic forces drive arteriogenesis, namely, increased luminal shear stress induces capillary expansion without branching and external stimulus induces vessel sprouting (Milner 2014; Ribatti 2015).

After recruitment of mural cells, especially smooth muscle cells, mural cells will sprout and migrate alongside the pre-existing vessels, accordingly to local hemodynamic forces. Arteries will be earlier and more extensively covered with smooth muscle cells (SMCs) than veins. Simultaneously, the smooth muscle cells differentiation is happening and these cells acquire special proprieties, such as contractility (Milner 2014; Ribatti 2015).

Additionally, significant remodeling of the vessels occur adapting to the blood flow changes of the target organ. Vessel remodeling processes occurs throughout all development and includes the growth of new vessels, the regression of others and establishing of new vessel network patterns. This process appears to be triggered and mediated by local environmental factors rather than genetically determined. Hemodynamic forces are also critical for reshaping the vascular networks, such as the type and angles of vessel branching (Milner 2014; Ribatti 2015).

In the early embryonic phases, there is no distinction between arteries and veins, which are morphologically similar. The differentiation is determined at a molecular level; different signalling pathways stimulate the differentiation into arteries or veins, such as ephrins, i.e., transmembrane ligands. This molecular explanation replaces the previous theory that hemodynamic forces were ruling the artery/vein differentiation (Milner 2014; Ribatti 2015).

Specifically regarding brain vasculature development, different steps have been identified, the most important being:

- First 2 weeks: The neural tissue receives their oxygen and nutrients through direct diffusion from the amniotic fluid.
- Between the second and forth week: Developing of the meninx primitive (mesodermal origin structure that is the forerunner of the meninges).
- The meninx primitive comprises a primitive perineural vascular plexus.

- The vasculogenesis starts with angioblasts migration into the head creating superficial extracerebral perineural vascular plexus, covering the brain. Several vascular maturation processes, such as extramural cell recruitment, vessel remodeling and regression, allow the development of meninx primitive meningeal vessels/plexus.
- These meningeal plexus receive the blood coming from the dorsal aortas and drain into the anterior cardinal veins. The anterior cardinal veins are the forerunners of the internal jugular veins. These are two short embryonic veins that, after confluence with the posterior cardinal veins forming the common cardinal veins, empty into the *sinus venosus* (forerunner of the atrium).
- Later, progressive intracerebral vessel sprouting angiogenesis occurs into the deeper neuroectodermal tissue towards the subventricular zone. These new vessels are initially located around the ventricles. The primitive choroid plexus result from the invagination of the neural tube, associated with the formation of the choroidal vessels.
- Subsequently, capillary development occurs with an inside-out order, extending from the ventricular surface into the developing white matter and cortical plate. The brain is supplied peripherally by the capillary network developing from the *meninx primitiva* vessels and deeply through the choroid plexuses.

During all these maturation steps, the brain vascularization (arterial and venous) development follows the metabolic needs of the emerging brain areas (Milner 2014; Ribatti 2015).

Intracranial Arterial Development

The carotid arterial system originates from the maturation of the aortic arches, also known as brachial/pharyngeal arches. From the aortic sac arise a pair of ventral aortas that communicate with the ipsilateral dorsal aorta through six paired aortic arches. The aortic are present simultaneously and they have complex development changes, including complete regression.

The first aortic arch (AA) is present at the 1.3 mm stage and successively; from cranial to caudal, the other aortic arches will develop (second AA at 3–4 mm; third and fourth AA at 4 mm; sixth AA at 5–7 mm stages). The fifth AA is not discussed in this chapter, since its existence, as a separate structure, and development is subject to on going controversy. Therefore, the aortic arch system is present from a very early stage (1.3 mm) until the 28 mm stage with regression of the sixth AA. A post-aortic arch phase with continuous remodeling of these arteries originates the definitive aortic arch, pulmonary, carotid, and subclavian arteries (Lasjaunias et al. 2001; Raybaud 2010).

The sequential arterial developments include (Fig. 1a–c):

- Dorsal aorta:
  - The pair of two arteries – dorsal aortas – present at the 4 mm stage will fuse into a single channel – aorta – at the 10 mm stage.
  - The dorsal aorta between the III and IV AA (ductus caroticus) regresses.
- 6th AA: Originates the pulmonary arteries.
- 5th AA: Regresses completely.
- 4th AA: The left 4th AA originates the aortic arch in association with the ventral aorta, and the right 4th AA forms the proximal subclavian artery.
- 3th AA: Proximal 3rd AA originates the common carotid artery, and the distal third AA segment together with the near dorsal aorta originate the internal carotid artery.
- 2nd AA: The regression of the 2st AA originates the hyoid artery (dorsal remnant), the carotid tympanic artery (ventral remnant), and a segment of the ventral remnant and the ventral aorta originate the ventral-pharyngeal artery.
- 1st AA: The regression of the 1st AA originates the supraorbital artery (dorsal remnant) and the mandibular artery (ventral remnant).

The external carotid artery embryonic development will not be addressed in this section, but the main external carotid artery branches progress from the following primitive arterial systems (Fig. 1e):

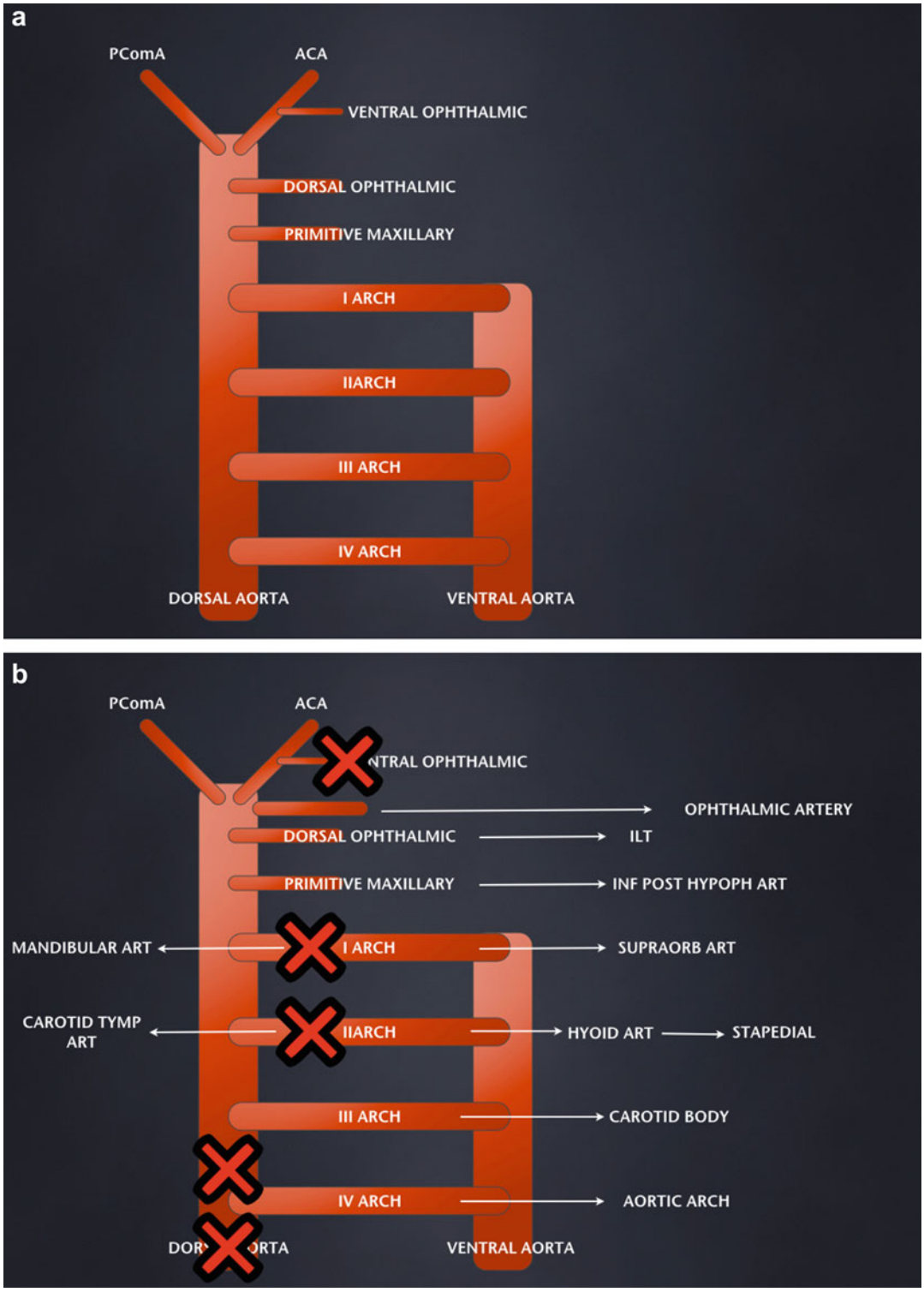


Fig. 1 (continued)

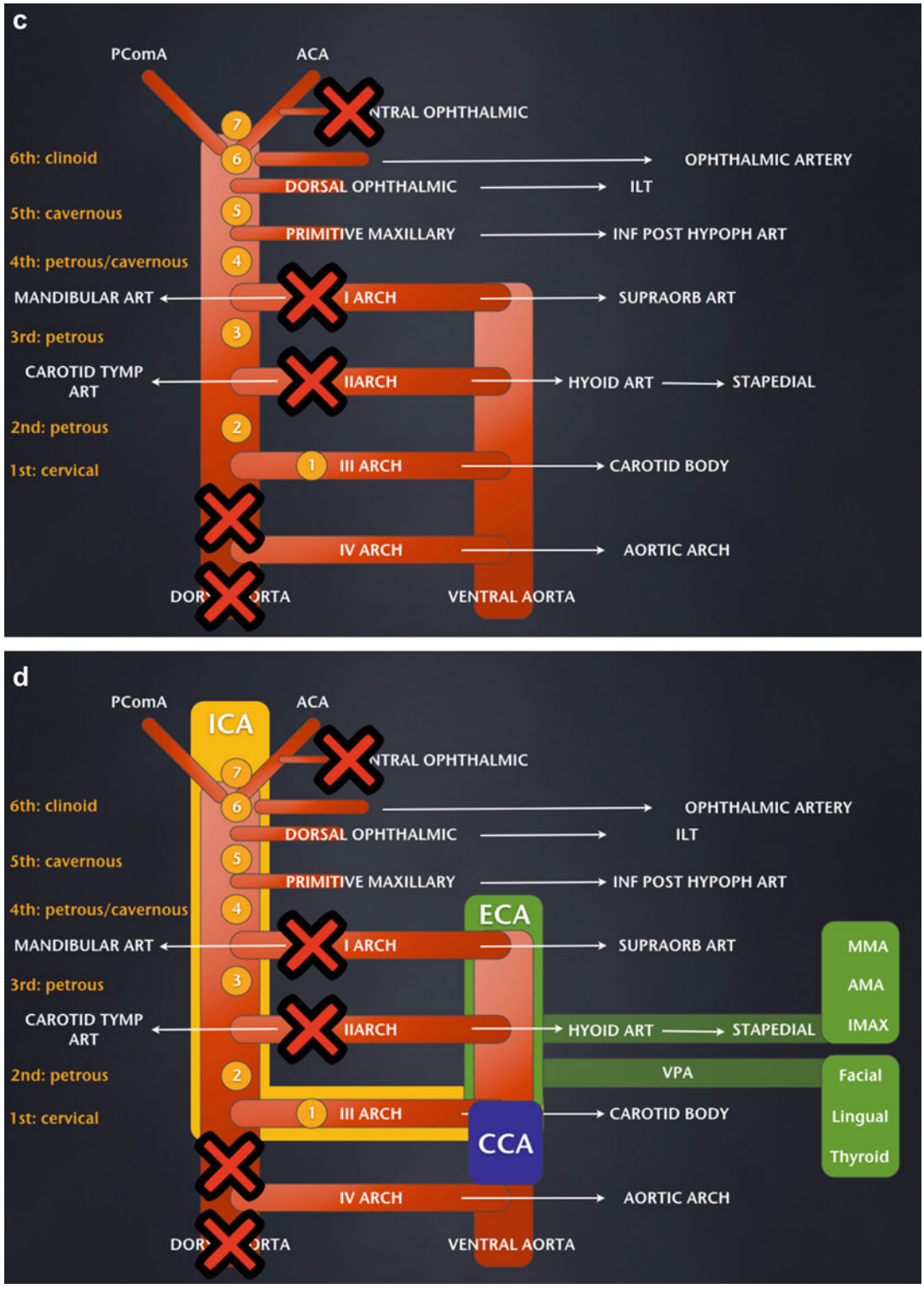


Fig. 1 (continued)

- The hyoid artery derived from the 2nd AA will generate the internal maxillary artery system.
- The ventral pharyngeal artery, originating mainly from the regressed ventral aorta will create the thyroid-facial-lingual artery system.
- The carotid-vertebro-basilar anastomosis (hypoglossal, proatlantal I and II) will originate the ascending pharyngeal-occipital artery system.

The internal carotid arteries (ICA) emerge at the 3 mm embryo stage resulting from the fusion of the 3rd AA and distal dorsal aorta. The distal embryonic ICA bifurcation occurs at the level of the posterior communicating artery and becomes evident at the 4 mm stage with the development of the anterior and posterior division (Lasjaunias et al. 2001; Raybaud 2010).

- The anterior (rostral) division: Includes the anterior cerebral artery and the anterior choroidal artery (and later the middle cerebral artery)
- The posterior (caudal) division: Includes the posterior communicating, posterior cerebral artery, superior cerebellar, and the posterior choroidal arteries

Initially, the anterior division is mainly responsible for the arterial supply of the optic and olfactory areas. Two major branches are identified: the medial olfactory artery, the forerunner of the anterior cerebral artery, and the lateral olfactory artery, which provides the striate (which originate Heubner artery and MCA) and posterior telencephalic (which will originate the anterior choroidal artery) arteries (Lasjaunias et al. 2001; Raybaud 2010).

The anterior cerebral artery originates from medial olfactory artery is the oldest cerebral cortical artery. The middle cerebral artery (MCA) is a recent phylogenetic acquisition resulting from the need to supply a large cortical territory of progressive enlarging brain. At the 11–12 mm embryological stage, it is possible to identify the MCA origin from the proximal anterior division of the ICA (i.e., MCA corresponds to an dominant – primitive – ACA perforator that annexes cortical territory). Initially, the "MCA" has a plexiform structure that will progressively fuse into a single channel at the 18 mm stage (Lasjaunias et al. 2001; Raybaud 2010).

The posterior division of the ICA will be responsible for the arterial supply of the occipital lobe and cerebellum through the posterior cerebral and superior cerebellar arteries and will fuse with the primitive vertebro-basilar system (originating from the parallel longitudinal neural channels).

Fig. 1 (**a**–**d**) Schematic representation of the evolution of the aortic arches, dorsal and ventral aortas at the head and neck region, and the correspondent arteries that originate from these embryonic vessels. (**a**) At the cranial end of the dorsal aorta is represented the embryonic internal carotid artery division: caudal division – PComA (posterior communicating artery); and anterior division – ACA (anterior cerebral artery). Dorsal ophthalmic artery; primitive maxillary artery; I-IV AA – aortic arches representation. The adult ophthalmic artery is the result of the maturation of three embryonic arteries: the ventral ophthalmic (originating from the ACA), the dorsal ophthalmic originating from the cavernous ICA segment (corresponding to the ILT), and the most common configuration in which the ophthalmic artery originates from the 6th ICA segment. (**b**) The dorsal ophthalmic artery will originate the inferolateral trunk (ILT). The primitive maxillary artery will originate the inferior and posterior hypophyseal artery. I aortic arch originate the mandibular artery and the supraorbital artery. II aortic arch originates carotid tympanic artery and the hyoid/stapedial arteries. The III aortic arch corresponds to the level of the carotid bulb. The IV aortic arch originates the adult aortic arch. (**c**, **d**) Internal carotid artery (1–7) embryonic segments (yellow) divided by the landmarks of the origin of embryonic arteries. CCA – common carotid artery (blue) and ECA – external carotid artery origins (green). The hyoid/stapedial system gives rise to the middle meningeal (MMA), accessory meningeal (AMA), and internal maxillary (IMAX) arteries. The thyroid-facial-lingual artery system (superior thyroid, lingual, and facial arteries) derives from the ventral pharyngeal artery (VPA) originating from the ventral aorta. Note: Although it is classically described that ITL is the forerunner of the primitive dorsal ophthalmic artery, and that an ophthalmic artery arising from ICA cavernous segment is named "dorsal ophthalmic artery" (variant), it is probably more precise to accept that the branch giving rise to the ILT is the primitive maxillary artery. The ophthalmic artery would be the result of the contribution of several embryonic arteries, namely: primitive maxillary artery from ICA cavernous segment, primitive dorsal and ventral ophthalmic arteries arising from the supraclinoid ICA segment, and stapedial artery and primitive olfactory artery from ACA

The circle of Wills is formed at the 6–7 weeks embryological stage with the development of the anterior communicating artery and "basilar tip" maturation (Lasjaunias et al. 2001; Raybaud 2010).

- The anterior communicating artery results from the medial extension of the anterior cerebral arteries, with a plexiform arterial structure that fuses at the midline at the 21–24 mm stage.
- The maturation of the posterior division of the ICA includes the cranio-caudal fusion with the basilar artery segment (originating from the longitudinal neural arteries), and midline fusion forming the distal end of the basilar artery ("basilar tip").

At an early stage (4–5 mm embryo), there are two parallel longitudinal neural channels corresponding to the cranial extension of the metameric arterial supply of the spine and spinal cord. From these parallel neural arterial channels arise several (transversal) segmental arteries (such as C1 or proatlantal 1 and C2 or proatlantal 2 arteries) that have a metameric distribution and follow the peripheral/cranial nerves (Lasjaunias et al. 2001; Raybaud 2010).

The hindbrain arterial supply is, initially and transiently, dependent of the carotid system, which comes from the transient carotid-vertebro-basilar (ventral-dorsal) anastomoses. These embryonic arterial anastomoses develop at a 5 mm or earlier embryo stage starting their involution at the 7–12 mm embryo stage. With the progressive enlargement of the posterior fossa structures, the carotid system becomes insufficient to provide adequate blood supply, and the posterior circulation system development is triggered. The carotid-vertebro-basilar (ventral-dorsal) anastomoses regression is inversely parallel to the development of the posterior circulation. If they persist, the proximal posterior circulation will be proportionally underdeveloped (hypoplastic). The life span of these anastomoses is extremely short, around 1 week, and complete regression occurs when the posterior communicating artery connects with the basilar artery segment arising from the ventral longitudinal neural arteries (Lasjaunias et al. 2001; Raybaud 2010).

The basilar artery is formed at an early stage (5–6 mm) by a complex process characterized by double fusion: midline fusion between the two parallel neural arterial channels; and a cranial-caudal fusion between the caudal ICA division and the midline fused parallel neural channels. It is classically accepted that the cranial-caudal fusion area corresponds to the level of the trigeminal artery (Lasjaunias et al. 2001; Raybaud 2010).

The vertebral arteries are originated from the intersegment arterial anastomosis between the cervical segmental arteries that occurs during the 7–12 mm stages, extending in a cranial-caudal fashion from the proatlantal system to the 6th cervical segmental artery (Lasjaunias et al. 2001; Raybaud 2010).

The cerebellar arteries originate from the posterior/caudal ICA division (Superior Cerebellar Artery – SCA) and from the development of the two parallel neural arterial channels (Anterior-inferior Cerebellar arteries – AICA and Posterior-inferior Cerebellar arteries – PICA). In analogy to the spinal cord arterial anatomy, AICA may be regarded as a dominant perforator (ventral/radiculo-medullar) that has annexed other territories such cortical cerebellar territory and PICA may be an analogue to a radiculo-pial (lateral/dorsal) spinal cord artery that has annexed other territories, such as cerebellum and choroid plexus (Lasjaunias et al. 2001; Raybaud 2010).

The carotid-vertebrobasilar anastomoses are: trigeminal artery, hypoglossal artery, proatlantal I and proatlantal II. The optic artery probably does not exist and the reported cases may represent a low (caudal) origin of the trigeminal artery (Fig. 2) (Lasjaunias et al. 2001; Raybaud 2010).

The trigeminal artery follows the V cranial nerve route, being responsible for its arterial blood supply. The persistent trigeminal artery is the most common carotid-vertebro-basilar anastomoses persistence with a prevalence of 0.1–0.6% approximately (Lasjaunias et al. 2001; Raybaud 2010).

The trigeminal arteries (TA) are generally subdivided into (Saltzman Classification):

- Type 1 – TA joins the BA between the superior cerebellar artery (SCA) and the anterior

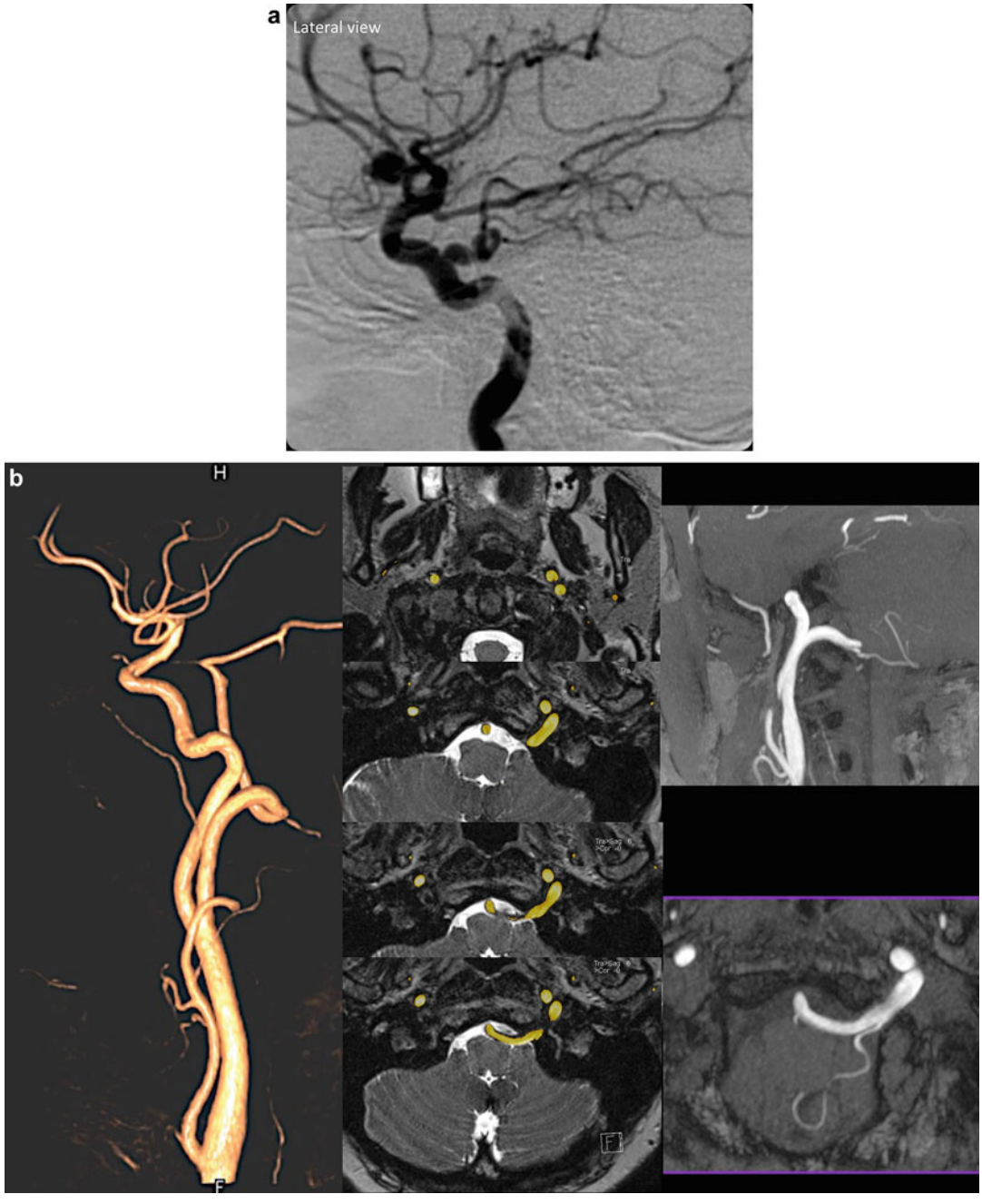


Fig. 2 (**a**, **b**) Carotid-vertebrobasilar anastomoses. (**a**) DSA lateral view after the right ICA contrast injection illustrates the presence of a trigeminal artery (TA) arising from the cavernous segment of the internal carotid artery (ICA) (caudal to the origin of the posterior communicating (PCommA)) and anterior choroidal artery (AChA) also seen in the angiogram. (**b**) 3D TOF VR and MIP images and fused high-resolution axial T2 and 3D TOF MRA showing hypoglossal artery persistence and the course of this artery at the hypoglossal canal

inferior cerebellar artery (AICA), the caudal segment of the BA can be hypoplastic.
- Type 2 – TA joins the BA above the origin of the SCA, and it supplies most of the flow to the SCA. Usually accompanied by normal PCOMM supplying most of the PCA flow.
- Type 3 – TA supply one of the cerebellar arteries ("persistent TA variant") bypassing the BA, was firstly described for SCA.

The Saltzman classification revision, subdivided the type 3 into:

- Type 3a – ICA gives rise to SCA
- Type 3b (the most common) – ICA gives rise to AICA
- Type 3c – ICA gives rise to PICA

The hypoglossal artery is the forerunner of ascending pharyngeal artery. It is a metameric artery that follows the course of the hypoglossal nerve, being responsible for its arterial blood supply. The persistent primitive hypoglossal artery originates at the cervical ICA, ascends parallel to the cervical ICA and enters the cranial cavity through the hypoglossal canal anastomosing with the caudal basilar artery segment (Lasjaunias et al. 2001; Raybaud 2010).

The proatlantal arteries correspond to the first cervical (C1) and second (C2) segmental and metameric cervical arteries. They follow the route of the C1 and C2 cervical nerves, respectively. The regression originates the C1 and C2 occipital and vertebral arteries branches. The persistence of proatlantal arteries is better classified accordingly to the route at the craniovertebral junction rather than from the ECA/ICA origin (Lasjaunias et al. 2001; Raybaud 2010).

- The type 1 proatlantal artery is the commonest proatlantal artery type, corresponding to approximately 60% of cases. It originates from the proximal ICA or ECA, ascends posterolaterally to the ICA and passes through the foramen magnum to join the ipsilateral intracranial vertebral artery.
- The type 2 proatlantal artery originates from the ECA, ascends posterolaterally to the ICA, passes through the transverse foramen of C1 vertebra to join the ipsilateral extracranial vertebral artery below the C1.

In the spine vascular metameric organization, each segmental artery receives the name of the nerve that it is following and supplies the peripheral nervous system (peripheral nerve), which is derived from the neural crest; the central nervous system (myelomere), which is derived from the neural tube; and the somite that has a mesodermic origin and is composed by dermis (dermatome), muscle (myotome), and bone (sclerotome). At the head, face, and cranio-vertebral junction areas, a similar metameric organization is present, which explains the phenotype appearance of the segmental vascular syndromes, which will be discussed later (Lasjaunias et al. 2001; Raybaud 2010).

Intracranial Venous Development

The venous drainage is composed of deep medullary and dorsal nuclear veins draining into the subependymal venous system and cortical medullary veins and ventral nuclear veins draining into the leptomeningeal venous system. There are also transcerebral/transmedullary veins anastomosis between the two systems (Lasjaunias et al. 2001; Raybaud 2010).

At the 4 mm embryo stage, it is possible to identify the earliest precursor of the intracranial venous system, a unique middle line located vein, *Vena Capitis Medialis* (or primary head vein). Afterwards, this vein will split into two major veins, which migrate laterally forming the primitive cranial sinuses (Lasjaunias et al. 2001; Raybaud 2010).

In the initial phases, during the weeks 6 to 8 (10–26 mm stage), the choroidal plexuses, the forerunner of the subependymal venous system, are formed. These plexuses are supplied by the ACA, AChA, and PChA and drain into dorsal choroidal veins that drain into a major venous

collector, the Markowski's vein (also denominated "primary internal cerebral vein" and "median prosencephalic vein"). This is a single vein, median and dorsal prosencephalic vein, that drains exclusively the blood from the choroid plexuses. The Markowski's vein appears at week 6 and regresses after the week 11, when the subependymal venous system, internal cerebral veins and the Galen vein have developed. The persistence of Markowski's vein is seen in association with Vein of Galen Aneurysmal Malformation.

At the 14 mm embryo stage, there are three major meningeal plexus (anterior, middle, and posterior) covering the developing brain and collecting their blood into the primitive cranial sinuses. These venous plexuses are in close relationship with the developing forebrain – prosencephalon (anterior venous plexus), midbrain – mesencephalon (middle venous plexus), and hindbrain – rhomboencephalon (posterior venous plexus). The primitive cranial sinuses drain their blood into the anterior cardinal vein, which is the forerunner of the internal jugular vein.

Between the 14 mm and 40 mm embryo stage, several venous developments occur: some primitive cranial sinuses regress to become the tentorial sinuses; the venous anastomosis (venous stalk) between the anterior and middle venous plexus will become the transverse sinuses, and the venous anastomosis (venous stalk) between middle and posterior venous plexuses will originate the sigmoid sinus. Additionally, between 24 and 40 mm embryo stage, there will be a middle line fusion of the venous plexus originating the superior sagittal sinus.

From the 3 months of fetal age (60–80 mm) until the postnatal development, the dural sinuses suffer significant remodeling processes. Transverse sinuses increase in diameter until of the 6 months of fetal age, acquiring a balloon shape. Then, after 7 months (fetal) until 2 years after birth (postnatal), they will decrease in diameter. Additionally, sigmoid sinuses will increase in diameter until the age of 2 years (postnatal).

The superior petrosal sinuses are late venous development acquisitions, occurring at the 80 mm embryo stage, deriving from the metencephalic veins connection with the cavernous sinuses.

The cavernous "sinuses" maturation is an important biological process since it has implication in intracranial venous drainage. Immediately after birth, all intracranial venous drainage depends fully on the sigmoid sinuses drainage. The use of the cavernous sinuses for brain venous drainage ("cavernous sinuses capture") occurs after birth until the age of 2 years through the connection of the cavernous "sinuses" with the superficial sylvian veins through the sphenoparietal sinuses.

The venous system not fully developed after delivery is an important feature of intracranial venous physiology. The post birth vessel maturation in one of the typical particularities of venous development and includes the cavernous sinuses maturation ("capture of the cavernous sinus"), the regression in size/disappearance of the occipital–marginal sinuses and the decrease in size of transverse sinuses.

Metameric (Segmental) Vascular Syndromes

The concepts of tissue embryological origin and precursor cell (cephalic neural crest and mesoderm cell) migration and differentiation explain some of the phenotypic appearance of complex vascular syndromes, metameric syndromes. If there is mutation of the neural crest or adjacent cephalic mesoderm before the cellular migration, it is expected to produce metameric malformations, with segmental distribution and different type of tissues involved.

These metameric diseases are defined as Spine and Craniofacial Metameric Syndromes. In the spine, this distribution is transverse, but at head and neck this distribution in less obvious. These syndromes may affect one or more metameres and may be bilateral. The type of vascular phenotypic expression may target mainly the capillary-venous tree in Cerebrofacial Arteriovenous Metameric Syndromes (CAMS) and Spinal Arteriovenous Metameric Syndromes (SAMS), the venolymphatic tree in Cerebrofacial Venous Metameric Syndromes (CVMS) and the arterial-capillary tree in PHACES (Posterior cranial fossa malformations,

Hemangiomas, Arterial anomalies, Coarctation of the aorta and cardiac defects, and abnormalities of the Eye) (Lasjaunias et al. 2001, 2006; Raybaud 2010).

The spine metameres are divided from 1 to 31 corresponding to the myelomeres. Lasjaunias et al. suggested that craniofacial metameres division into: CMS type 1 with a midline topographic distribution, medial prosencephalic (olfactory) group, involving the hypothalamus, corpus callosum, hypophysis, and nose; CMS type 2 with lateral topographic distribution, prosencephalic (optic) group, involving the optic nerve, retina, thalamus, parietal, temporal, and occipital lobes and maxilla; CMS type 3 with caudal distribution, rhombencephalic (otic) group, involving of the cerebellum, pons, petrous bone, and mandible (Lasjaunias et al. 2001, 2006; Raybaud 2010).

The Cobb's syndrome (SAMS) is characterized by multiple vascular malformations (AVM) affecting different tissues of a myelomere, namely, the spinal cord, nerve root, bone, paraspinal space, subcutaneous, and skin.

The Bonnet-Dechaume-Blanc or Wyburn-Mason syndrome (CAMS) presents with multiple, high-flow AVMs having a metameric distribution affecting the brain, orbit (retinal or retrobulbar lesions), and maxillofacial region (Lasjaunias et al. 2001, 2006; Raybaud 2010).

The Sturge-Weber syndrome is the typical example of CVMS. It generally affects the first two metameres (CVMS 1–2) and is characterized by cortical/pial venous obstructions with collateral venous circulation through capillary venous proliferation and enlargement of the transmedullary collateral venous drainage, with or without choroid plexus hypertrophy. Other common features include facial "port wine stains," lymphangiomatous malformation, and facial/skull base bone hypertrophy. Secondary brain abnormalities, such as gyral calcifications and atrophy, are commonly seen as the result of chronic pial venous occlusion (Lasjaunias et al. 2001, 2006; Raybaud 2010).

The typical finding in PHACES is the presence of a large facial hemangioma. Intracranial hemangiomas are also seen. Intracranial arterial stenoses have been also added to the syndrome spectrum. The phenotypic expression of this disease is more complex affecting the arterial-capillary vascular tree and multiple metameres. This causes different types of phenotypic lesion expression and distinct target lesions, such as hemangiomas and arterial abnormalities and intracranial and thoracic localization. It may be explained by a larger and longitudinal cephalic neural crest dysfunction (Lasjaunias et al. 2001, 2006; Raybaud 2010).

Intracranial Arterial Anatomy: Anterior and Posterior Arterial Circulation

The intracranial arterial circulation is generally divided into two major territories: the anterior circulation, encompassing the internal carotid artery circulation (internal carotid (ICA), anterior choroidal (AChA), anterior cerebral (ACA), and middle cerebral arteries (MCA), and the posterior circulation comprehending the vertebro-basilar system (vertebral arteries, basilar artery, postero-inferior cerebellar arteries (PICA), antero-superior cerebellar arteries (AICA), superior cerebellar arteries (SCA), and posterior cerebral arteries (PCA).

Reminding the embryonic development, the primitive ICA anterior (cranial) embryonic division corresponds to the anterior circulation; and the primitive ICA posterior (caudal) embryonic division and the vertebro-basilar system correspond to the posterior circulation.

The brain arteries have superficial/cortical branches that are responsible for the arterial supply of the brain surface and deep/perforator branches that supply the brain stem, grey nuclei, and deep white matter.

The superficial/cortical arteries are responsible for the supply of the brain surface, including cortex and adjacent subcortical white matter. They have prominent leptomeningeal anastomosis at the brain surface. Additionally, the supratentorial cortical arteries are connected at the base of the brain through the circle of Willis.

The ACA, MCA, and PCA are responsible for arterial supply of the cerebrum.

- ACA supplies the medial surface of the frontal and parietal lobes, the inferior surface of the

frontal lobe, the cingulate gyrus, and corpus callosum.
- MCA supplies the lateral surface of the frontal, parietal, and temporal lobes.
- PCA is responsible for the supply of the temporal, occipital, and parietal lobes.

The SCA, AICA, and PICA (cerebellar arteries) are responsible for arterial supply of the cerebellum.

- SCA supplies the superior (tentorial) cerebellar surface.
- AICA supplies the anterior (petrosal) cerebellar surface.
- PICA supplies the inferior (occipital) cerebellar surface.

The perforators (deep arteries) are responsible for the arterial supply of the inner brain structures, namely, the thalamus, basal ganglia, and deep white matter, cerebellar nuclei, and the brain stem.

The perforators arising from the anterior circulation (ICA, AChA, ACA, ACM) and posterior cerebral artery (PCA) may be grouped into the:

- Anterior choroidal group, encompassing the ICA and AChA perforators
- Striatal group, including the medial and lateral lenticulostriate arteries originating from the ACA and MCA
- Thalamic group, comprising posterior choroidal arteries, the thalamoperforators, thalamogeniculate, thalamotuberal arteries arising from the posterior communicating artery, PCA, and basilar tip

The posterior circulation perforators arising from the basilar artery, vertebral arteries, and cerebellar arteries (SCA, AICA, PICA) are responsible for the arterial supply of the brain stem (medulla and pons).

Anterior Circulation

The anterior circulation is composed by the internal carotid artery (ICA) system, including the anterior and middle cerebral arteries and the anterior choroidal arteries. The posterior communicating artery branches will be discussed together with the posterior cerebral artery anatomy. The proximal ICA branches, namely, the petrous and cavernous segment branches and the ophthalmic artery are beyond the scope of this chapter.

Internal Carotid Artery Segment (ICA) – Intradural Segment

Different classifications have divided the internal carotid artery (ICA) into several embryological and/or topography segments.

Accordingly, to the embryological development, Lasjaunias et al. proposed the segmentation of the ICA into seven segments (Lasjaunias et al. 2001):

- 1st segment – cervical segment: From the carotid artery bifurcation into the ICA entrance at the skull base
- 2nd segment – ascending petrous segment: Starts at the carotid foramen and extends to the origin of the carotico-tympanic artery at the petrous bone
- 3rd segment – horizontal petrous segment: In-between the origins of the carotico-tympanic artery and vidian/mandibular artery, crossing the carotid canal until the foramen lacerum
- 4th segment – ascending cavernous segment: Extends within the cavernous sinus from the origins of the vidian/mandibular artery and the meningo-hypophyseal trunk
- 5th segment – horizontal cavernous segment: Extends inside the cavernous sinus from the origin of the meningo-hypophyseal trunk into the infero-lateral trunk origin
- 6th segment – clinoidal segment: Extends in-between the origins of infero-lateral trunk and ophthalmic artery
- 7th segment – terminal/supraclinoid segment: Extends from ophthalmic artery and anterior cerebral artery having an intradural course.

A surgical anatomical classification considers four ICA segments:

- C1 segment (cervical portion): Extending from the carotid bifurcation to the skull base

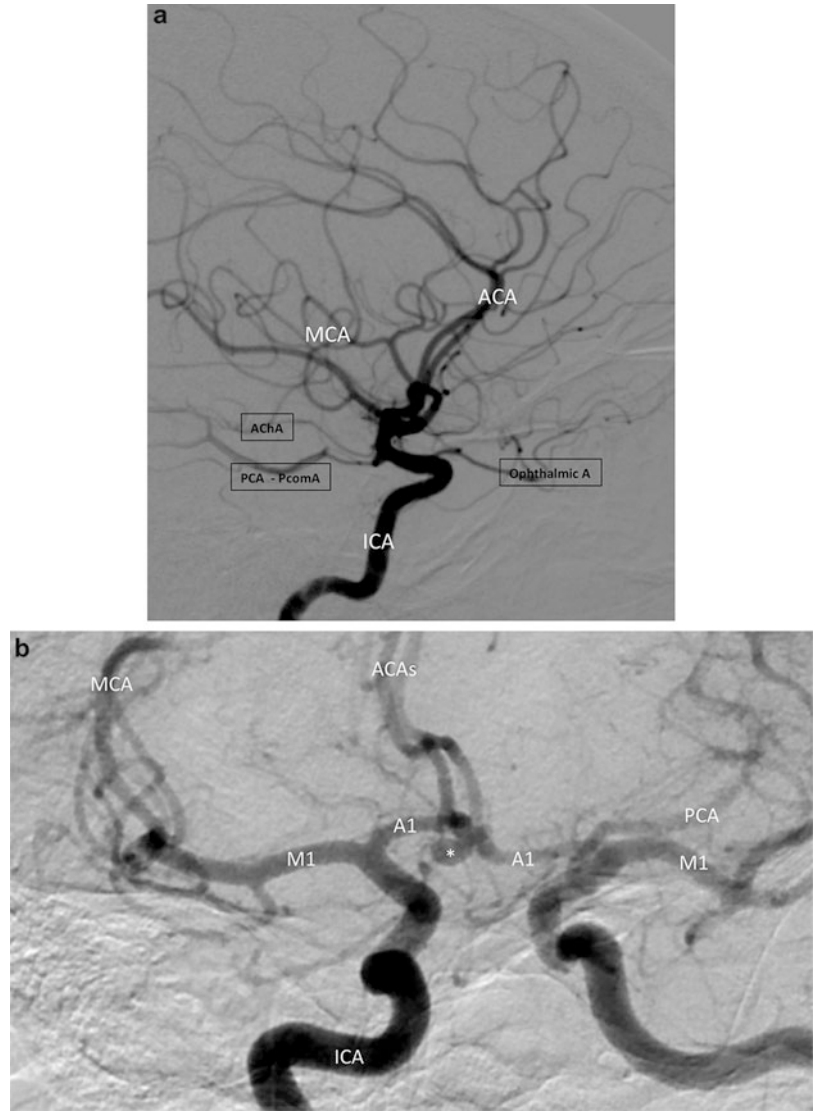


Fig. 3 (**a**, **b**) Internal carotid artery (ICA). (**a**) DSA lateral view after the right ICA contrast injection illustrates the major branches of the supraclinoid ICA segment: posterior communicating, anterior Choroidal (AChA), middle cerebral (MCA), and anterior cerebral arteries (ACA). (**b**) DSA ap/frontal view after the bilateral ICA contrast injection illustrates the proximal branches of the supraclinoid ICA segment and the proximal segments of ACA and MCA in a patient with an anterior communicating aneurism (*)

- C2 segment (petrous): Running within the carotid canal and terminating at the cavernous sinus entrance
- C3 segment (cavernous): Coursing inside the cavernous sinus
- C4 segment (supraclinoid): Corresponding to the intradural artery segment ending at the bifurcation into the anterior and middle cerebral arteries

The intradural ICA gives rise to the ophthalmic, posterior communicating and anterior choroidal arteries and has several perforator branches (Fig. 3).

The proximal group of perforators, arising near the ophthalmic artery, include the superior hypophyseal arteries (supplying the pituitary stalk and gland) and small branches to the optic chiasm and nerves and the floor of the III ventricle. There is a small group of perforators originating from the segment near the posterior communicating artery origin supplying the optic chiasm/tract, the floor of the III ventricle and the infundibulum. The ICA segment near the origin of the anterior choroidal artery has the largest number of perforators and supply the optic track; the infundibulum and the anterior and posterior perforate substances.

Anterior Choroidal Artery (AChA)

The anterior choroidal artery is one of the oldest (phylogenetic) arteries being of primordial importance for the brain supply during the embryonic choroidal stage. During embryonic development, AChA supplies a significant cortical (telencephalic) territory that will be later annexed by the posterior cerebral artery. Therefore, there is an inverse relationship between the sizes of AChA and PCA. In some rare congenital variants, a large cortical territory may remain under the supply from AChA, with a concordant posterior cerebral artery hypoplasia (Lasjaunias et al. 2001; Raybaud 2010).

In adults, AChA originates from the supraclinoid (intradural) ICA segment distally to the posterior communicating artery origin, at the carotid cistern, runs through the perimesencephalic cistern (cisternal segment), enters the choroidal fissure and branches at the level of the temporal horn choroid plexus (plexal segment) (Lasjaunias et al. 2001; Rhoton 2002c).

The cisternal segment of the AchA supplies the optic tracts, cerebral peduncles, posterior limb of the internal capsule, globus pallidus, lateral geniculate body, and mesial temporal lobe (hippocampus, dentate gyrus, and fornix). For these territories, there is a balance between the deep territory of the AChA and the branches from ICA, PCA, PComA, and MCA. The plexal segment gives branches to the choroid plexus of the temporal horn, atrium, and body of the lateral ventricles (Lasjaunias et al. 2001; Rhoton 2002c).

Anterior Cerebral Artery (ACA)

The anterior cerebral artery arises from the distal supraclinoid ICA segment, coursing medially, generally above the optic chiasm and nerves, entering the interhemispheric fissure (Figs. 4a and b).

The anterior cerebral artery is divided into five anatomical segments, namely (Rhoton 2002c):

- A1 (precommunicating segment): Corresponding to the segment between ICA and Anterior Communicating Artery
- A2 (infracallosal segment): Corresponding to the segment between the rostrum and the genu of the corpus callosum
- A3 (precallosal segment): Corresponding to curve around the genu of the corpus calosum
- A4 (supracallosal segment): Corresponding to the segment that follows the superior part of the corpus callosum and the "coronal suture" point
- A5 (posterocallosal segment): Corresponding to the segment located distal to the "coronal suture point"

ACA can exhibit several anatomical variants, specially comprising the A2 segment and the pericallosal and calloso-marginal arteries, but the distribution of the deep and cortical branches is generally constant. Most commonly, the A2 segment gives rise to the orbito-frontal and polar frontal arteries and at the distal end of the A2 segment the ACA divides into: pericallosal artery and calloso-marginal artery. The sizes of pericallosal and calloso-marginal arteries are in balance and vary inversely (Lasjaunias et al. 2001; Rhoton 2002c).

The pericallosal artery runs at the corpus callosum sulcus giving branches to the corpus callosum and distally originating the precuneal artery and small branches at the level of the splenium of the corpus callosum. The later, anastomose with the corresponding branches of the posterior cerebral artery (posterior pericallosal artery) (Lasjaunias et al. 2001; Rhoton 2002c).

The calloso-marginal artery courses at the cingulate sulcus and giving rise to the medial (anterior, middle, and posterior) frontal arteries and paracentral artery (Lasjaunias et al. 2001; Rhoton 2002c).

The internal/medial frontal arteries can arise from the calloso-marginal artery or from the A2 segment (anterior medial frontal artery) and pericallosal artery (middle medial frontal artery). The paracentral artery originates from the calloso-marginal artery (Lasjaunias et al. 2001; Rhoton 2002c).

Some anatomical variants may occur at the distal end of the ACA system, being in balance with the PCA vascularization. ACA at the A4/A5

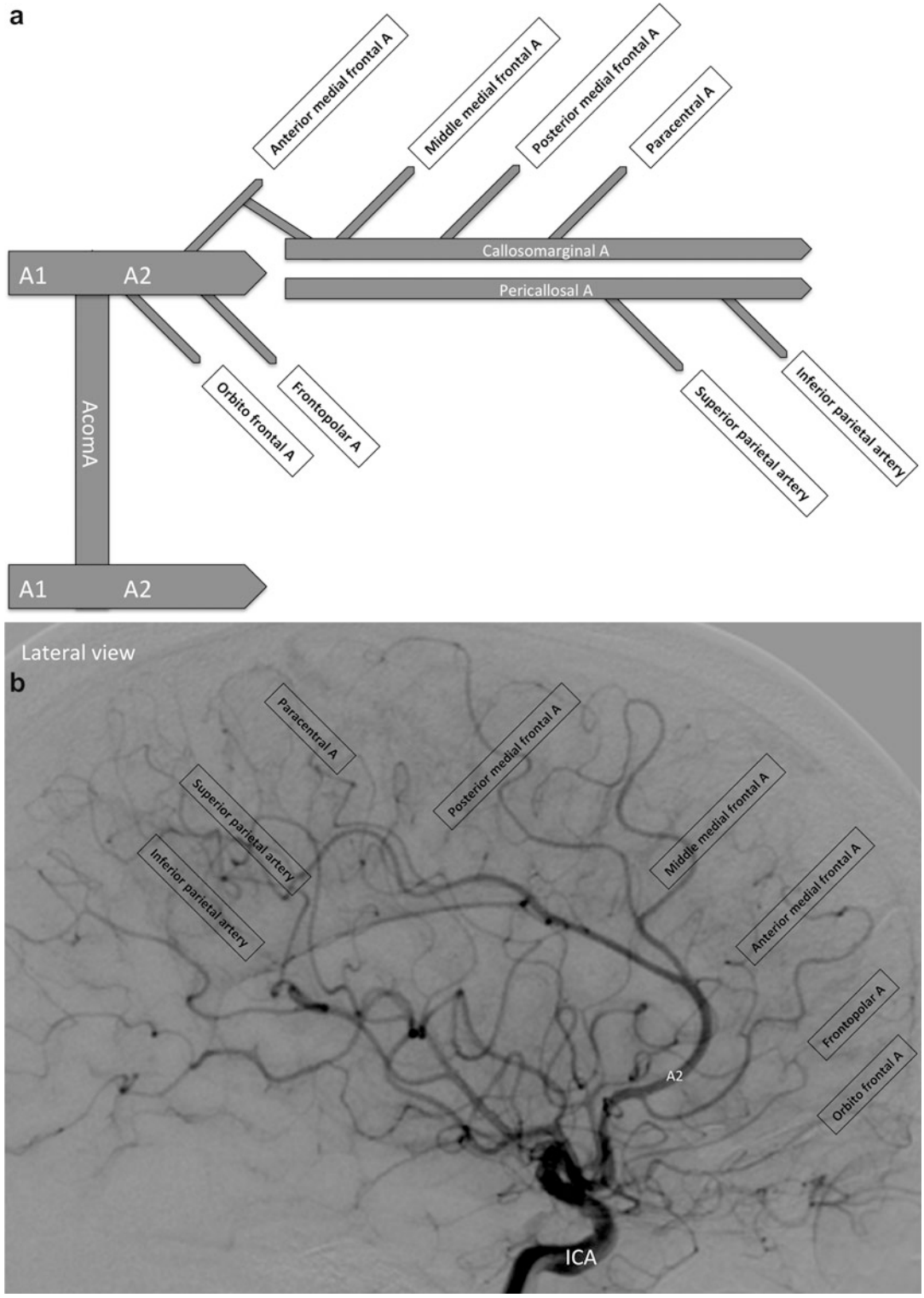


Fig. 4 (**a**, **b**) Anterior cerebral artery. (**a**) Schematic representation of the ACA branching pattern. (**b**) Anterior cerebral artery. DSA lateral view after the right ICA contrast injection illustrates the ACA cortical branches

segment of the calloso-marginal may give rise to superior (precuneus) and inferior parietal arteries and inferior callosal artery (Lasjaunias et al. 2001; Rhoton 2002c).

The major ACA cortical branches include:

- Orbito-frontal/medial fronto-basal artery
- Polar frontal artery
- Medial anterior frontal artery
- Medial intermediate frontal artery
- Medial posterior frontal artery
- Paracentral artery
- Parietal arteries: Superior (precuneal) artery; inferior parietal artery

The deep (perforators) branches from the ACA system arise most commonly from the anterior communicating artery and the A1 ACA segment.

Anterior communicating artery: It gives rise to hypothalamic branches, optic nerve/chiasm branches and the subcallosal artery that supply the septum pellucidum, paraterminal gyrus, subcallosal area; anterior commissure; fornix, lamina terminalis (Lasjaunias et al. 2001; Rhoton 2002c).

A1 ACA segment: It gives rise to the medial lenticulostriate arteries. The lateral (initial) half of the A1 segment has more perforators than the medial (distal) half. The recurrent artery of Heubner is the largest ACA perforator, originating at the A1, Acomm-A1 junction, being responsible for the supply of the head of the caudate nucleus and anteroinferior portion of the anterior limb of the internal capsule (other supply includes anterior third of the putamen, anterior part of the outer segment of the *globus pallidu*s, uncinate *fasciculus*) (Lasjaunias et al. 2001; Rhoton 2002c).

Middle Cerebral Artery (MCA)

MCA is a late embryonic acquisition and the largest cortical cerebral artery. It is responsible for the arterial supply of large cerebral cortical and deep brain territories (Fig. 5a, b, and c).

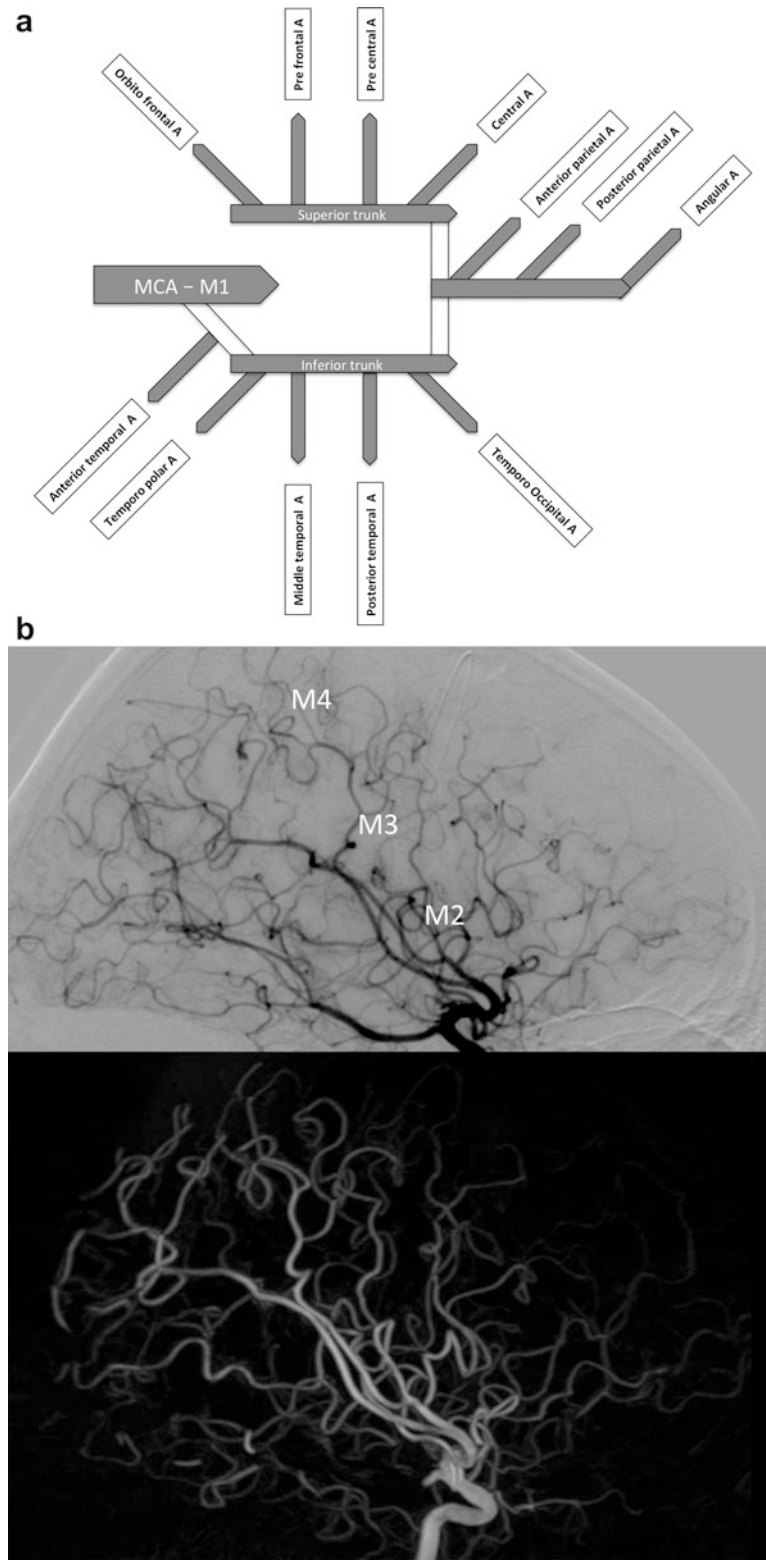
The middle cerebral artery is divided into four anatomical segments, namely (Lasjaunias et al. 2001; Rhoton 2002c):

- M1 (Sphenoidal segment): Starts at the distal ICA segment, extending laterally within the Sylvian fissure until the MCA division; small cortical branches (such as anterior temporal artery) may arise from this segment;
- M2 (Insular segment): This segment includes the MCA division trunks that lie in front of the insula and give rise to short cortical branches to the insula;
- M3 (Opercular segment): Begins at the insula circular sulcus and ends at the surface of the sylvian fissure and is responsive for the cortical branches to the operculum;
- M4 (Cortical segment): Corresponds to the cortical branches of the MCA.

There is a wide variation of MCA division and dominance of the MCA trunks, but the cortical arteries distribution is fairly constant. The MCA may divide into two major trunks ("bifurcation"), the superior and the inferior trunks, of a similar size (co-dominance). There may be an inferior or superior dominance with a larger territory supplied by one of the trunks. Other possibilities are the presence of three major MCA division trunks ("trifurcation") or multiple (more than three) trunks (Lasjaunias et al. 2001; Rhoton 2002c).

The major M4 (cortical) branches include (Lasjaunias et al. 2001; Rhoton 2002c):

- Lateral frontobasal/orbitofrontal artery
- Prefrontal artery
- Precentral (pre-rolandic) artery
- Central (rolandic) sulcus artery
- Anterior parietal (postcentral sulcus) artery
- Posterior parietal artery
- Angular artery
- Temporooccipital artery
- Posterior temporal artery
- Intermediate temporal artery
- Anterior temporal and temporopolar arteries



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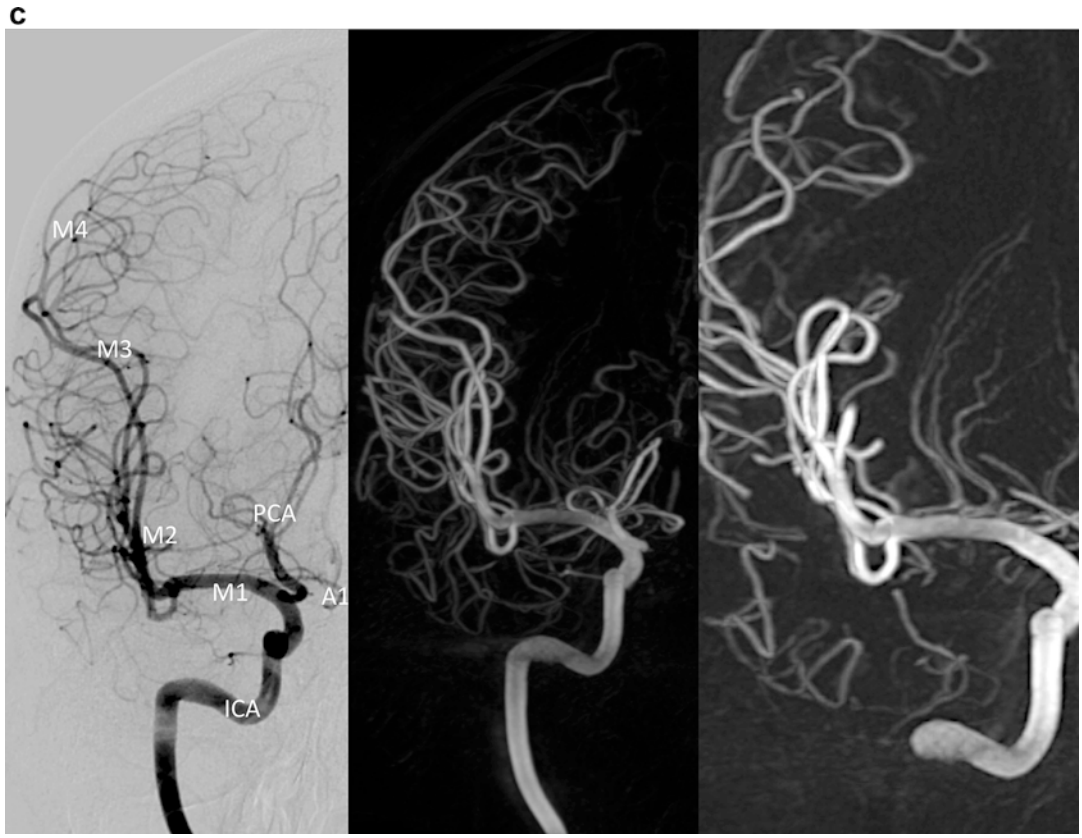


Fig. 5 (**a**–**c**) Middle cerebral artery. (**a**) Schematic representation of the MCA branching pattern. (**b**) DSA lateral view after the right ICA contrast injection (2D image – upper image; 3D MIP image – lower image). (**c**) Anteroposterior view after the right ICA contrast injection (2D image – right image; 3D MIP image – middle and left images – show the different MCA segments, the origin of the perforators (lenticulostriate arteries) from the M1 segment and the cortical branching pattern

The group of lateral frontobasal/orbitofrontal artery, prefrontal artery, precentral (pre-rolandic) artery, and central (rolandic) sulcus artery arises generally from the superior MCA trunk.

The group of temporo-occipital artery, posterior temporal artery, intermediate temporal artery, commonly originates from the inferior MCA trunk (Lasjaunias et al. 2001; Rhoton 2002c).

The anterior temporal and temporo-polar arteries can arise from the M1 segment or from the inferior trunk (Lasjaunias et al. 2001; Rhoton 2002c).

The origin of the anterior parietal (postcentral sulcus) artery, posterior parietal artery, and angular artery is more variable: Balanced origin from superior and inferior trunks (co-dominance); origin from one of the trunks (inferior/superior trunk dominance); origin from an individual trunk ("trifurcation") (Lasjaunias et al. 2001; Rhoton 2002c).

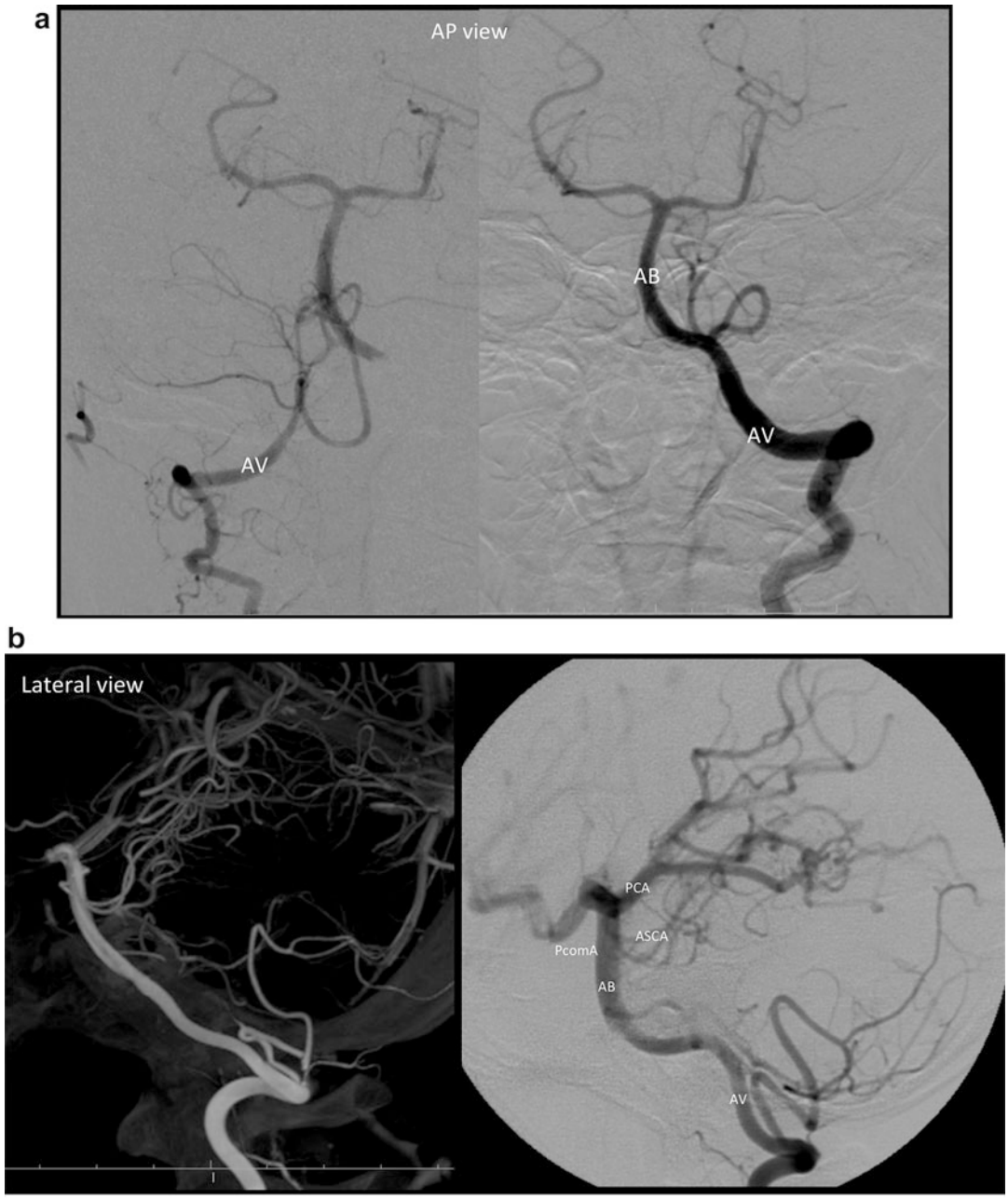
The deep (perforators) branches from the MCA system arise mostly (up to 80% of the total branches) from the M1/prebifurcation segment.

The perforator branches of the MCA are the lenticulo-striate arteries, which may be subdivided into (Lasjaunias et al. 2001; Rhoton 2002c):

- Medial lenticulo-striate arteries which are an inconstant group of 1 to 5 arteries that may not be present and originate from the initial M1 (prebifurcation) segment.
- Intermediate lenticulostriate arteries originating from the M1 (prebifurcation) segment.
- Lateral lenticulo-striate arteries that are the most constant group, which can arise from the M1 or M2 segments.

Posterior Circulation

The vertebro-basilar system constitutes the posterior circulation. The posterior circulation has two embryonic origins: from the internal carotid artery (ICA) system, comprising the posterior cerebral artery and superior cerebellar artery (originating from the caudal division of the "embryonic" ICA); and from the metameric organized arterial system, encompassing the vertebral arteries, PICA, and AICA (Fig. 6a, b, and c) (Lasjaunias et al. 2001).



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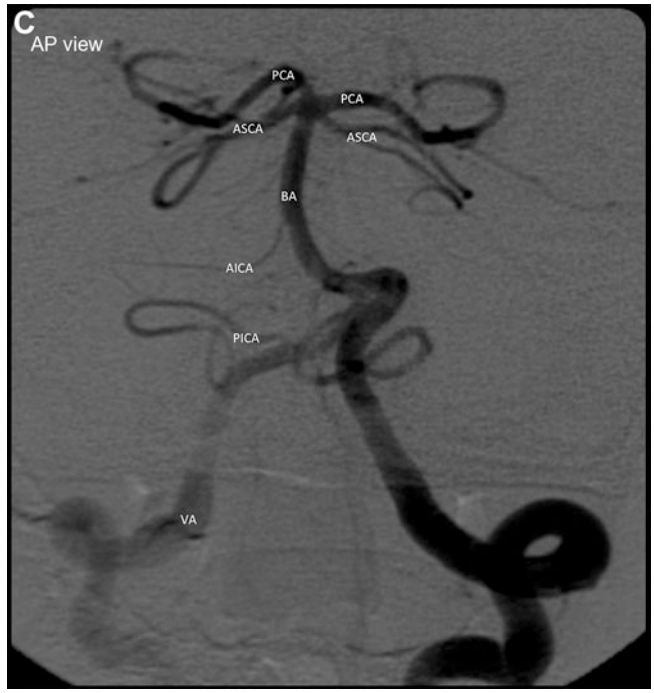


Fig. 6 (**a**–**c**) Vertebral and basilar arteries. (**a**) DSA anteroposterior view after right VA (right image) and left VA (left image) contrast injection illustrates the course and branches of the VAs and BA and the presence of right VA hypoplasia mostly at the V4 segment. (**b**) DSA lateral view after VA contrast injection (3D MIP image – right image; 2D image – left image) illustrates the course and branches of the VAs and BA and the filling of the posterior communicating artery (PComA) on the left image. (**c**) DSA anteroposterior view after left VA contrast injection shows the origins and courses of the cerebellar arteries and posterior cerebral arteries

Vertebral and Basilar Arteries

The vertebral arteries originate at the level of the subclavian arteries, having an ascending route within the neural foramen and entering the cranial cavity through the foramen magnum. They are generally divided into four segments (Lasjaunias et al. 2001):

- V1 (extradural/extracranial segment extending from the subclavian artery to the entrance at the 6th transverse foramina)
- V2 (extradural/extracranial segment extending from 6th to 2nd transverse foramina
- V3 (extradural segment extending from the second transverse foramina to the foramen magnum crossing the atlas transverse foramina, the upper surface of the posterior atlas arch and ascending to the foramen magum)
- V4 (extradural/intracranial segment ascending at the anterolateral medullary surface to join the contralateral and continue as the basilar artery)

At the cervical segments, the vertebral arteries give rise to several small muscular and radicular/spinal branches. The intracranial vertebral arteries branches are meningeal branches, spinal branches, such as the posterior spinal and/or anterior spinal arteries, several perforating branches for the brain stem, and the posterior inferior cerebellar artery (PICA).

The basilar artery starts with the vertebral arteries confluence at the level of the ponto-medullary junction, running grossly at the mid-line of the basilar sulcus (central pontine sulcus), within the prepontine cistern ventral to the pons, until the interpeduncular cistern at the level of ponto-mesencephalic junction. At the interpeduncular

cistern, the basilar tip divides into two pairs of arteries, the superior cerebellar and posterior cerebral arteries. The basilar artery provides supply for both the infratentorial and supratentorial territories. Along its course, the basilar artery gives rise to several perforators for the brain stem and the antero-inferior cerebellar artery (AICA) (Lasjaunias et al. 2001).

Posterior Communicating Artery (PCommA)

The posterior /caudal ICA division is the forerunner of the posterior communicating artery (PcommA). The PCommA arises from the póstero-lateral surface of the ICA supraclinoid segment, running backwards (oblique trajectory: posterior/ascending and medial) within carotid and interpeduncular cisterns, joining the posterior cerebral artery. PCommA has several perforator branches, namely, the premammilar artery (which is an anterior thalamo-perforating artery), the thalamo-tuberal and thalamo-perforating arteries, supplying hypothalamus, anterior thalamus, posterior limb of the internal capsule, tuber cinereum, mammilary bodies, subthalamic nucleus, and posterior perforated substance (Duvernoy 2010; Lasjaunias et al. 2001; Rhoton 2002c).

Posterior Cerebral Artery (PCA)

Posterior cerebral artery begins at the basilar tip and is in communication with the carotid system through the posterior communicating artery (Fig. 7).

Anatomically, the posterior cerebral artery is generally divided into four segments, namely (Lasjaunias et al. 2001; Rhoton 2002c):

- P1 (precommunicating) segment: Corresponds to the initial segment between the basilar tip and the posterior communicating artery.
- P2 (crural and ambient) segment: Running in the crural and ambiens cisterns and ending at the postero-lateral margin of the midbrain. It

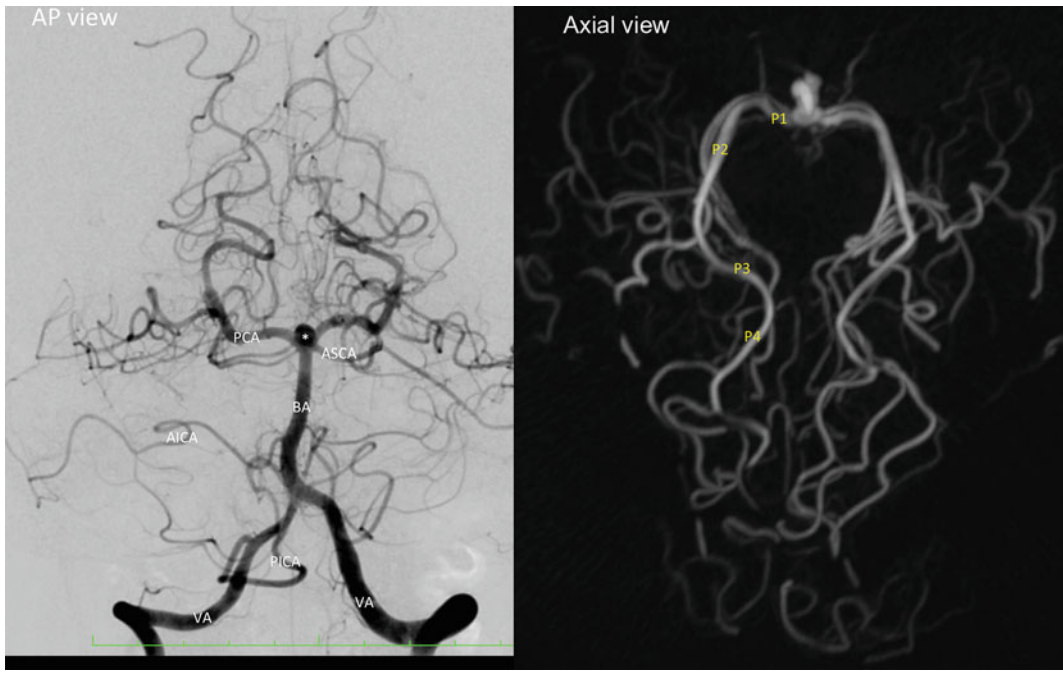


Fig. 7 Posterior cerebral artery. DSA anteroposterior view after VA injection (right image) and 3D MIP axial image obtained from 3D DSA, illustrates the PCA course and distal cortical branches

can be subdivided into P2 a segment (anterior: Crural) and P2 P segment (posterior: Ambient).
- P3 (quadrigeminal) segment: Coursing at the quadrigeminal cistern and ending at the calcarine fissure anterior limit. At the distal end of the P3 segment, PCA bifurcates into parietooccipital and calcarine arteries.
- P4 (cortical) segment: Corresponds to the distal cortical branches segment that begins at the end of calcarine sulcus.

The major PCA cortical branches include (Lasjaunias et al. 2001; Rhoton 2002c):

- Anterior temporal artery
- Posterior temporal artery
- Posterior (peri)callosal artery
- Lateral occipital artery
- Anterior/inferior temporal artery
- Middle/inferior temporal artery
- Posterior/inferior temporal artery
- Medial occipital artery
- Parietooccipital artery
- Calcarine artery

The deep/perforator branches of the PCA arise from the P1 and P2 segments and supply mainly the midbrain and diencephalon. From the P1 segment originate direct (short) and circumferential (short and long) perforating arteries and the thalamo-perforating arteries. From the P2 segment originate direct (short) and circumferential (short and long) perforating arteries, the thalamo-geniculate arteries, the medial posterior choroidal arteries (MPChA) – from the P2 A segment, and the lateral posterior choroidal arteries (LPChA) – from the P2 P segment; the latter can also originate from P3 segment (Duvernoy 2010; Lasjaunias et al. 2001; Rhoton 2002c).

- Direct (short) perforating arteries supply the cerebral peduncles and geniculate bodies. Circumferential (short and long) perforating arteries supply the quadrigeminal plate, pulvinal, and also cerebral peduncles.
- Thalamo-perforating arteries supply the diencephalic-mesencephalic junction, cerebral peduncles and mesencephalic tegmentum, medial-ventral thalamus, hypothalamus, and posterior limb internal capsule.
- Thalamo-geniculate arteries supply posterior lateral thalamus, posterior limb of the internal capsule, optic tract.
- The MPChA and LPChA supply the choroid plexus of the lateral ventricles but also give rise to the superior and posterior (pulvinar) thalamic arteries, respectively.
  - The MPChA also supplies the cerebral peduncles, tegmentum, geniculate bodies (medial and lateral, but primarily the former), the colliculi, pineal gland, pulvinar, and medial thalamus.
  - The LPChA also supplies cerebral peduncle, posterior commissure, part of the crura and body of the fornix, the lateral geniculate body, pulvinar, dorsomedial thalamic nucleus, and the body of the caudate nucleus.

Cerebellar Arteries: SCA, AICA, PICA

The cerebellar arteries – Superior Cerebellar Artery (SCA), Anterior-Inferior Cerebellar Artery (AICA), Posterior-Inferior Cerebellar Artery (PICA) – supply the cerebellum and the brain stem.

These three arteries may be divided into four anatomical segments (Rhoton 2000a):
- Antero-medial (cisternal) segment: This segment is in close relationship with the anterior brainstem surface: Anterior ponto-mesencephalic segment (SCA); anterior pontine segment (AICA); anterior medullary segment (PICA).
- Antero-lateral (cisternal) segment: This segment is in close relationship with the lateral brainstem surface: Lateral ponto-mesencephalic segment (SCA); lateral pontine segment (AICA); lateral medullary segment (PICA).
- Lateral (fissure) segment: This segment is in close topographic relation with the cerebello-brain stem fissures: Cerebello-mesencephalic

fissure (SCA), cerebello-pontine fissure (AICA) and cerebello-medullary fissure (PICA).

- Posterior (cortical) segment: This segment covers the superior/tentorial (SCA), anterior/petrosal (AICA), inferior/occipital (PICA) cerebellar cortex.

Superior Cerebellar Artery (SCA)

SCA is the most constant cerebellar artery originating at the basilar tip. Its origin can have single (~86% of cases), double (~14% of cases), or triple (<1% of cases) trunks (Lasjaunias et al. 2001; Rhoton 2000a).

The SCA brainstem perforators arise from the antero-medial and antero-lateral segments. From the lateral segment originates the pre-cerebellar arteries that are responsible for the arterial supply of the dentate nuclei, superior cerebellar peduncle, and inferior colliculi (Rhoton 2000a).

The SCA generally divides into a rostral medial (vermian) and caudal lateral (hemispheric) branch at the cisternal segments. The marginal artery arises from the rostral branch at the anterolateral segment and anastomosis with AICA.

At posterior (cortical) segment, the rostral branch subdivides distally the into medial and paramedical branches supplying the vermis. The caudal branch subdivides into lateral, intermediate, and medial hemispheric branches supplying the upper cerebellar hemisphere (Fig. 8) (Rhoton 2000a).

Anterior-Inferior Cerebellar Artery (AICA)

The AICA originates from the basilar artery (more frequently from the caudal half of the basilar artery) as (~72% of cases), double (~26% of cases) or even a triple (~2% of cases) trunks. Considering an AICA duplication/triplication these branches should supply cerebellar cortex, otherwise they should be considered as large perforators (Lasjaunias et al. 2001; Rhoton 2000a).

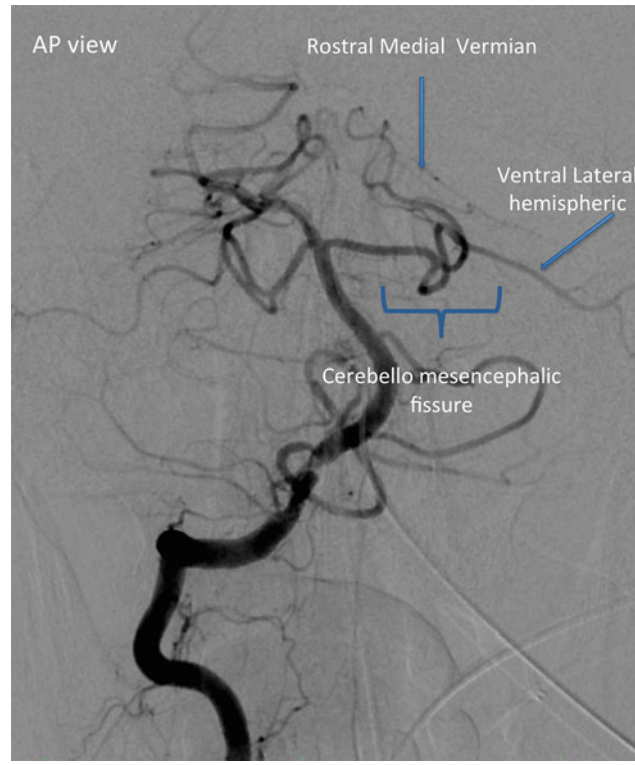


Fig. 8 Superior cerebellar artery. DSA anteroposterior view after right VA contrast injection illustrates the SCA course and cortical branches and specifies the segment at the cerebellomesencephalic fissure in which the branches that supply the dentate nucleus arise

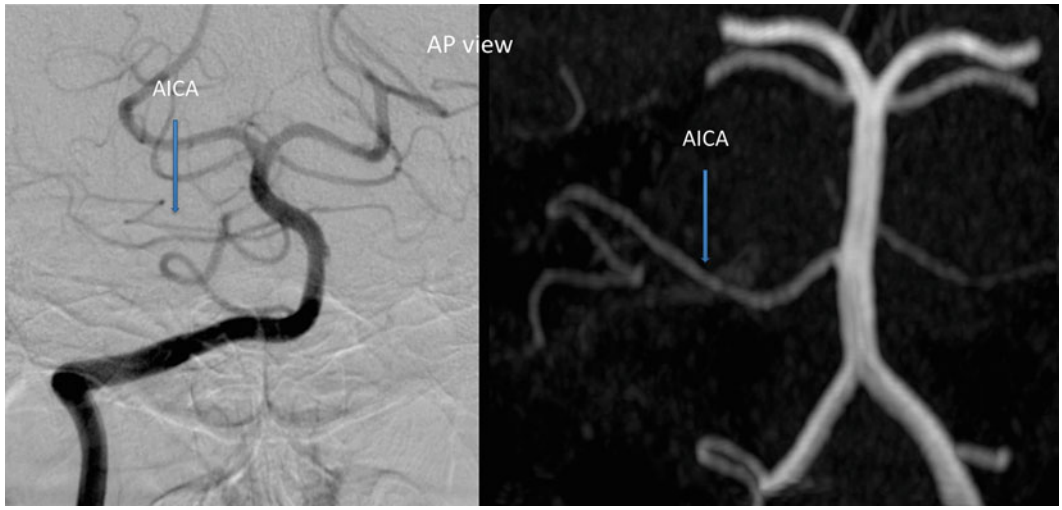


Fig. 9 Anteroinferior cerebellar artery. DSA anteroposterior view after right VA contrast injection (right image) and 3D TOF MRA MIP image (left image) exemplify the AICA course and branches, and identify the course at the internal auditory meatus (M-shape configuration) and illustrate a variant with AICA duplication

The perforators to the brain stem originate from the cisternal segments. The labyrintine artery to the inner ear and the subarcuate artery being responsible for the petrous bone dural supply and the anastomosis with the ascending pharyngeal and stylomastoid arteries originate from the AICA main trunk or the rostral trunk at the cisternal segment (Duvernoy 2010; Lasjaunias et al. 2001; Rhoton 2000a).

AICA main trunk subdivides into cortical branches that run over the petrous cerebellar surface, namely the rostral branch supplying the inferior cerebellar petrosal surface and the caudal branch supplying the inferior cerebellar petrosal surface, flocculus and IV ventricle choroid plexus (Fig. 9) (Duvernoy 2010; Lasjaunias et al. 2001; Rhoton 2000a).

Posterior-Inferior Cerebellar Artery (PICA)

The PICA is the most variable cerebellar artery, regarding its origin, territory and size. Most commonly, occurs as a single artery originating from the intradural (V4) vertebral artery (in up to 83% of cases). Rarely there is a PICA origin duplication (~3% of cases) or extradural origin (below the foramen magnum) (Lasjaunias et al. 2001; Rhoton 2000a).

The perforators to the brain stem arise from the cisternal segments. The cerebello-medullar segment exhibits a double curve: caudal loop (tonsillo-medullary segment) and cranial loop (telovelo-medullary segment). From cisternal and lateral segments choroidal arteries originate that supply the IV ventricle choroidal plexus and tela choroidea (Duvernoy 2010; Lasjaunias et al. 2001; Rhoton 2000a).

Generally, at the lateral segment, PICA subdivides into two cortical branches: a larger lateral hemispheric branch supplying the tonsills and the lateral inferior cerebellar hemispheric surface and a medial smaller vermian branch supplying the vermis (Fig. 10) (Lasjaunias et al. 2001; Rhoton 2000a).

Variant Anatomy of Willis Circle and Other Intracranial Vessels

The circle of Willis (CoW) is formed at a very early phase (20–40 mm stage). The CoW is composed of the A1 (ACA) segments, anterior communicating artery (ACommA), supraclinoid ICA segments, P1 (PCA) segments,

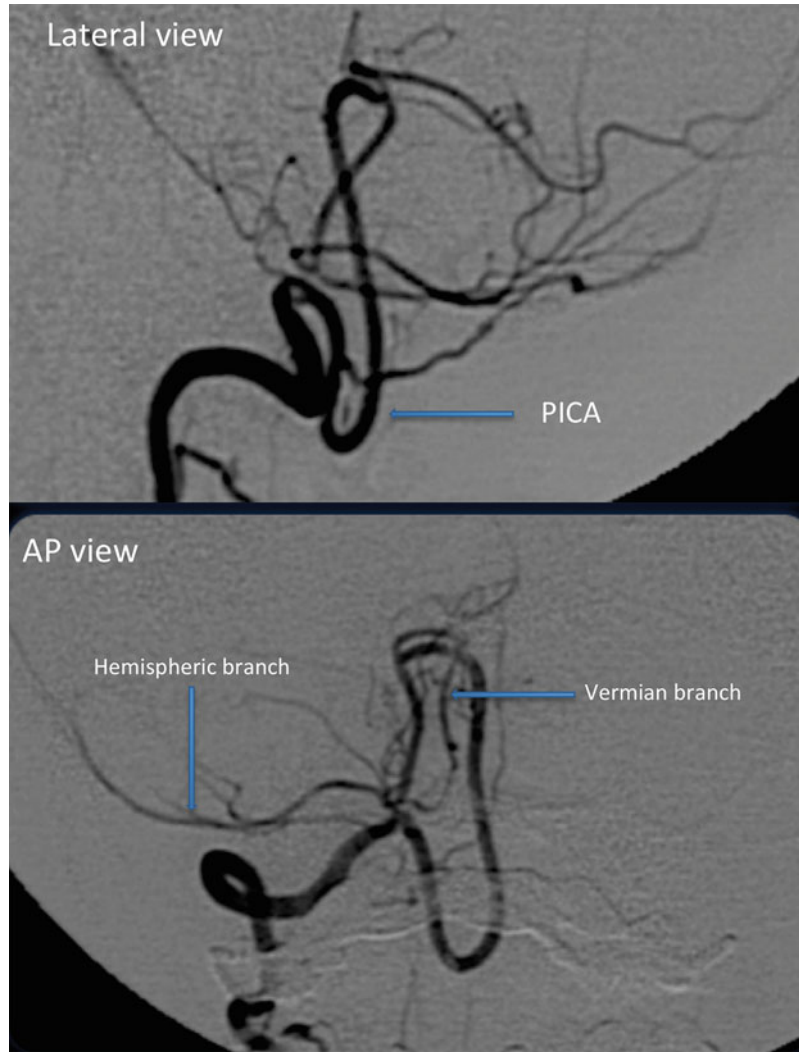


Fig. 10 Posteroinferior cerebellar artery. DSA lateral (upper image) and anteroposterior (lower image) views after right VA contrast injection illustrates the PICA course and cortical branches. This PICA has an extradural origin at the level of C1

posterior communicating arteries (PCommA) and basilar tip.

During the embryonic development of the CoW there are two major maturation steps (Lasjaunias et al. 2001; Raybaud 2010):

- Middle line plexiform fusion of the ACA medial extensions, which originates the anterior communicating artery
- Middle line fusion of the posterior/caudal division of the ICA originating the basilar tip

The congenital anatomical variants of the CoW are very common, with different type of size asymmetries and hypoplasia. Some of the most common examples is the segmental (A1, P1, PcomA) hypoplasia. P1 hypoplasia may be associated with an enlarged ipsilateral PComA in a pattern classically termed as "fetal" circulation type. The presence of ACommA fenestration represents the persistence embryonic appearance of these vessels that have a primitive plexiform structure.

CoW variations, such as hypoplasia or arterial segment absence, may explain different outcomes in acute ischemic stroke and/or unusual arterial stroke territorial distribution. Arterial morphological abnormalities, such as fenestrations, may also be associated with an increased risk for development of vascular disorders, like intracranial aneurysms (Lasjaunias et al. 2001).

Other common arterial variants include (Lasjaunias et al. 2001):

- Persistence of carotid-vertebro-basilar anastomosis (Lasjaunias et al. 2001)
  - Trigeminal artery
  - Hypoglossal artery
  - Proatlantal type 1
  - Proatlantal type 2
- Anterior cerebral artery (ACA)
  - Azigos artery: Fusion of the initial ACA segments into a single trunk that provides branches for both hemispheres
  - Bihemispheric ACA: One ACA supplies both hemispheres (associated with unilateral A1 hypoplasia)
  - Multiple (triplicated) ACA: Several separated branches arise from proximal ACA segments
  - Ventral ophthalmic artery: Ophthalmic artery origin from the ACA
  - Infraoptic course of ACA: A1 segment courses below the optic nerve and chiasm
- Middle cerebral artery (MCA)
  - MCA fenestration: Persistence of the plexiform primitive structure
  - Duplicated MCA: MCA is formed by two arterial trunks originating from the supraclinoid ICA, with the perforator branches arising from the distal trunk
  - Accessory MCA: MCA origin from A1 segment or ACommA (the perforator branches arise from the cranial trunk)
- Anteroinferior cerebellar artery (AICA)
  - AICA-PICA: Dominant AICA artery that annexes ipsilateral PICA territory
- Posteroinferior cerebellar artery (PICA)
  - PICA-AICA: Dominant PICA artery that annexes ipsilateral AICA territory
  - Bihemispheric PICA: PICA supplying both cerebellar hemispheres with a fusion of the vermian branches

Intracranial Venous Anatomy and Variability

Intracranial Venous Anatomy

The intracranial venous anatomy is organized into the superficial and deep venous system. The deep venous system drains into the vein of Galen and the superficial venous system drains through cortical veins into the dural sinuses and cavernous plexuses. Both systems are connected through the transmedullary veins in a hemodynamic equilibrium.

The dural sinuses are intradural venous structures, covered by the two dural layers, are in close relationship with dural attachments, such as those at the tentorium and falx cerebri. They have a membranous origin, similar to the dura of the cranial vault convexity. The dural sinuses are the superior sagittal, inferior sagittal, straight, transverse and sigmoid sinuses (Fig. 11a) (Rhoton 2000b, 2002a, b).

The venous plexus have an epidural location, between the periosteum and the dura. They have a cartilaginous origin, similar to the dura of the cranial base. These venous structures include the cavernous, retroclival venous plexuses, the venous plexus of the foramen magnum and the spine epidural venous plexuses (Fig. 12).

The supratentorial superficial venous system is composed of cortical veins that drain into four major venous collectors, namely: (Fig. 11b and c) (Duvernoy 1975; Rhoton 2002b)

- Superior sagittal group, draining into the superior sagittal sinus
- Sphenoid group converging into the sphenoparietal sinus or cavernous venous plexus
- Tentorial group, draining into the venous tentorial sinuses, located at the upper surface of the tentorium and emptying into the transverse sinuses
- Falcine group that drain into the inferior sagittal or straight sinus, or their tributaries

The superior sagittal sinus (SSS) has a variable length, with a variable of the anterior segments. The anterior third of the sinus may be absent. At the anterior extremity it may communicate with the nasal veins and with emissary veins through its entire course. The superior sagittal sinus ends at the torcular Herophili. The torcular Herophili receives also the blood from the straight sinus and continues through the transverses sinuses. The SSS receives the blood from several cortical veins from the superior part of the medial and

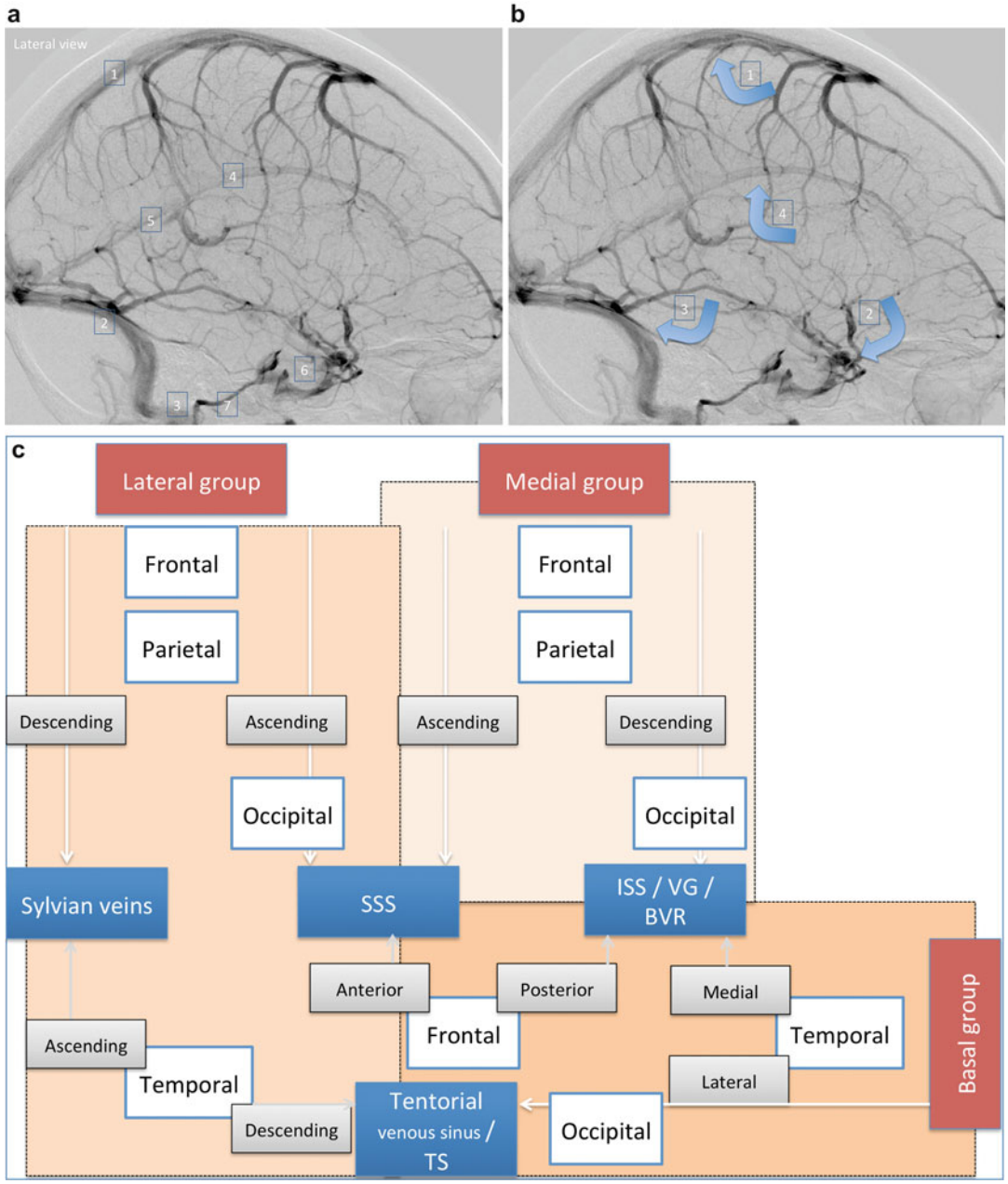


Fig. 11 (**a**–**c**) (**a**) Dural sinus anatomy. DSA venous phase lateral view after ICA contrast injection shows the major dural sinuses. (**b**) Dural sinus anatomy. DSA venous phase lateral view after ICA contrast injection shows the four major routes for superficial venous drainage (1-superior sagittal, 2-sphenoid 3-tentorial, and 4-falcine groups). (**c**) Schematic representation of the supratentorial cortical venous distribution and preferential drainage

lateral surfaces of the frontal, parietal, and occipital lobes and the anterior part of the orbital surface of the frontal lobe (Duvernoy 1975; Lasjaunias et al. 2001; Rhoton 2002b).

The transverse and sigmoid sinuses establish the communication between the torcular Herophili and the internal jugular vein. The transverse sinuses run within the tentorium attachment

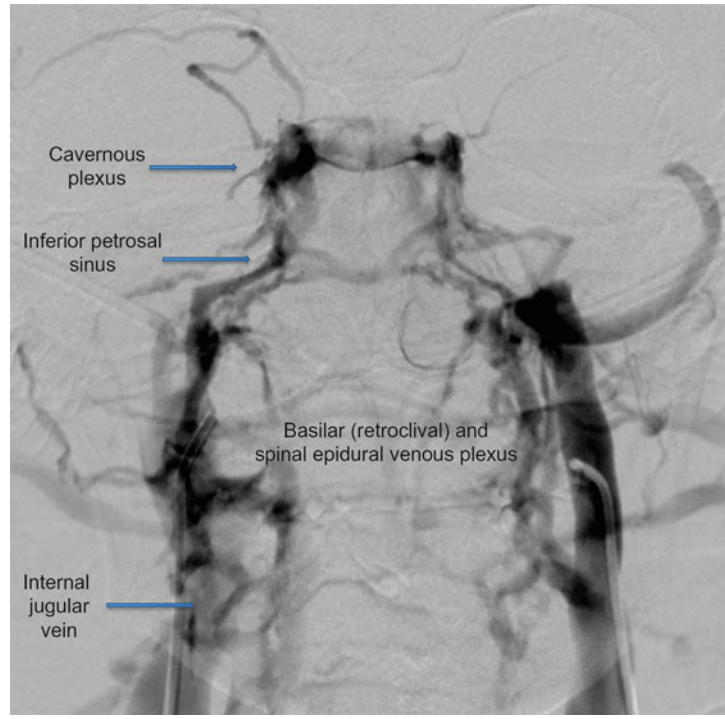


Fig. 12 Cavernous sinus plexus. DSA anteroposterior view after bilateral internal jugular vein contrast injection demonstrates the cavernous plexus and the venous connections with the inferior petrosal sinus, basilar plexus, and sylvian vein

until the origin of the superior petrosal sinus and the sigmoid sinus are positioned in-between the origins of the superior and inferior petrosal sinuses (Duvernoy 1975; Lasjaunias et al. 2001; Rhoton 2002b).

The transverse sinus drains in both supra- and infra-tentorial cortical veins. They receive the blood indirectly through tentorial sinuses located within the tentorium, the medial tentorial sinus draining the cerebellum and the lateral tentorial sinus draining the temporal and occipital lobes, receiving among others the vein of Labbé.

The sigmoid sinuses communicates with the inferior petrosal sinuses, does not generally receive cortical veins, but has relevant venous anastomoses with the suboccipital venous plexus, condylar veins, marginal and occipital sinus and cavernous plexus (through the inferior petrosal sinus) (Duvernoy 1975; Lasjaunias et al. 2001; Rhoton 2002b).

The inferior sagittal sinus (ISS) courses at the inferior edge of the falx cerebri ending at the vein of Galen – straight sinus junction. Along its course there are venous channels communicating with the superior sagittal sinus. It receives blood from the internal parietal, frontal lobe, and cingulate gyrus through medial descending internal veins (Duvernoy 1975; Lasjaunias et al. 2001; Rhoton 2002b).

The cavernous "sinuses" are epidural venous plexus located between the perosteum and the dura matter, draining the face-orbit and the brain, and constitute the cephalic extension of the spine and basilar (clival) epidural venous plexuses. As previously mentioned, the cavernous plexuses are important alternative intracranial venous drainages but they are only fully matured after birth, until around the second year of life (Rhoton 2002a, b).

The cavernous plexus are a venous hub at the level of the skull base, they have:

- Contralateral communication through the anterior and posterior inter-cavernous sinuses
- Posterior continuation with the inferior and superior petrosal sinuses and basilar plexus
- Inferior communication with the extracranial pterygoid plexuses through emissary veins

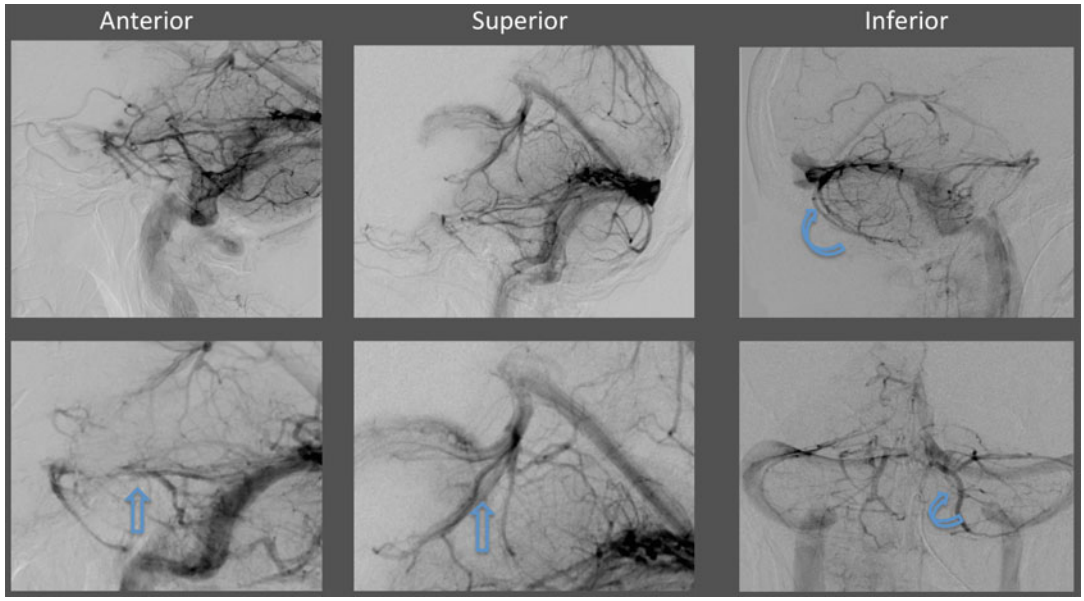


Fig. 13 Posterior fossa veins. DSA venous phase lateral view after ICA contrast injection shows the three major venous pathways: anterior to the petrous veins; superior to the vein of Galen, and inferior into the tentorial sinus and transverse sinus

- Anterior communication with the superior and inferior (or common venous confluent) ophthalmic veins
- Lateral communication with the sphenoparietal sinus and sylvian veins

The superior petrosal sinuses run above the petrous bone from the junction of the transverse and sigmoid sinuses and the cavernous plexus. It receives the blood from the posterior fossa anterior (petrosal) venous group, which is generally gathered by the petrosal veins. The inferior petrosal sinus runs at the petroclival fissure, does not receive cortical venous blood, and acts as a venous anastomotic pathway between the cavernous plexus and the internal jugular veins (Duvernoy 1975; Lasjaunias et al. 2001; Rhoton 2002b).

The straight sinus runs from the union of the inferior sagittal sinus and the vein of Galen to the torcular Herophili within a dural attachment of the falx cerebri and tentorium (Duvernoy 1975; Lasjaunias et al. 2001; Rhoton 2002b).

The infratentorial superficial venous system is composed of superficial cortical, deep, and brain stem veins (Fig. 13).

The infratentorial cortical veins drain into three major venous collectors, namely (Duvernoy 1975; Lasjaunias et al. 2001; Rhoton 2002b):

- Anterior (Petrosal) group: Draining into the petrosal vein(s) and/or superior petrosal sinuses
- Superior (Galenic) group: Emptying into the basal vein of Rosenthal and/or vein of Galen
- Inferior (Tentorial) group: Draining into the venous tentorial sinuses, located at the lower surface of the tentorium and empting into the transverse sinuses

The anterior (petrosal) group, drains the anterior cerebellar surface, includes the anterior (superior, middle and inferior) cerebellar hemispheric veins, gathered at the great horizontal fissure (Duvernoy 1975; Lasjaunias et al. 2001; Rhoton 2002b).

The superior vermian (anterior ascending) and superior hemispheric (anterior ascending) groups

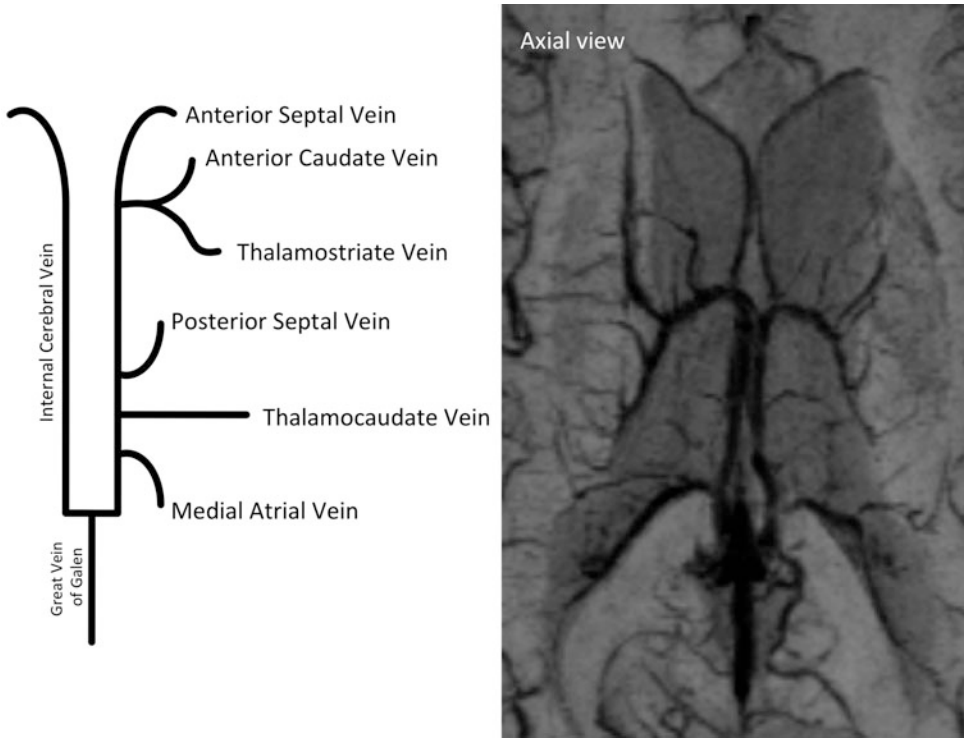


Fig. 14 Schematic representation of the deep venous system and corresponding axial SWI MRI images illustrating the veins draining into the internal cerebral veins. (Courtesy of Dr.ª Teresa Nunes – Portugal)

drain the superior cerebellar surface constitute the superior group (galenic) (Duvernoy 1975; Lasjaunias et al. 2001; Rhoton 2002b).

The inferior (tentorial) group drains the inferior cerebellar surface through the inferior vermian veins, inferior hemispheric veins, retro-tonsillar veins, medial and lateral tonsillar veins. The superior vermian (posterior descending) and superior hemispheric (posterior descending) veins also participate in this group (Duvernoy 1975; Lasjaunias et al. 2001; Rhoton 2002b).

The infratentorial brain stem and deep venous system is formed by an extensive venous network of veins with longitudinal and transversal arrangement draining into the vein of Galen, basal veins of Rosenthal and petrosal veins. They have caudal venous anastomoses with the spinal cord veins and the marginal sinus (Duvernoy 1975; Lasjaunias et al. 2001; Rhoton 2002b).

The supratentorial deep venous system is composed of the internal cerebral veins, basal vein of Rosenthal and their venous confluents, being responsible for the venous drainage of the midbrain, limbic lobe, corpus callosum, pineal gland, choroid plexuses, basal ganglia thalami and deep cerebral white matter (Duvernoy 1975; Lasjaunias et al. 2001; Rhoton 2002b).

The internal cerebral veins are formed by the confluence of the medial septal vein and the thalamo-striate veins at the level of the Monro foramina. This vein runs within the velum interpositum and at the level of the quadrigeminal cistern it joins the contralateral internal cerebral vein forming the vein of Galen (Fig. 14) (Duvernoy 1975; Lasjaunias et al. 2001).

Along its course, the internal cerebral veins receive several subependymal veins, which can be divided into two groups, namely, the:

- Lateral group – including the anterior and posterior septal veins and medial atrial veins (Duvernoy 1975; Lasjaunias et al. 2001)

- Medial group – including the anterior and posterior caudate veins, thalamo-striate veins, superior striate, and lateral atrial veins

The septal veins receive venous blood from medullary veins arising from the frontal lobe (anterior septal veins) and parietal/occipital lobes (posterior septal veins). The atrial veins drain the parietal/occipital lobe medullary veins (medial atrial veins) or the temporal /occipital lobe medullary veins (lateral atrial veins). The other medial group drains mainly the basal ganglia and thalamus, the internal capsule and the deep white matter (Duvernoy 1975; Lasjaunias et al. 2001).

The basal vein of Rosenthal receives blood supply from both superficial and deep veins. It is responsible for the cortical venous drainage of the medial and inferior brain surface, namely the medial temporal lobe, the orbital frontal lobe surface, and the insula. It also drains the hypothalamus, midbrain, striatum, internal capsule, and thalamus (Duvernoy 1975; Lasjaunias et al. 2001).

The basal vein of Rosenthal is generally divided into three segments (Duvernoy 1975; Lasjaunias et al. 2001; Rhoton 2002b):

- Anterior – corresponding to the segment from the origin at to the cerebral peduncle ventral surface. It receives the anterior cerebral and deep middle cerebral vein at its origin. Along its course it receives the inferior striate vein and the olfactive, insular, orbito-frontal, and uncal veins.
- Medial – corresponding to the segment located within the crural and ambiens cisterns. It receives the blood from the peduncular veins, hippocampal veins, and inferior ventricular and choroidal veins.
- Posterior – corresponding to the final segment that joins the vein of Galen or the internal cerebral vein. This is the most variable segment being even absent in some cases. In

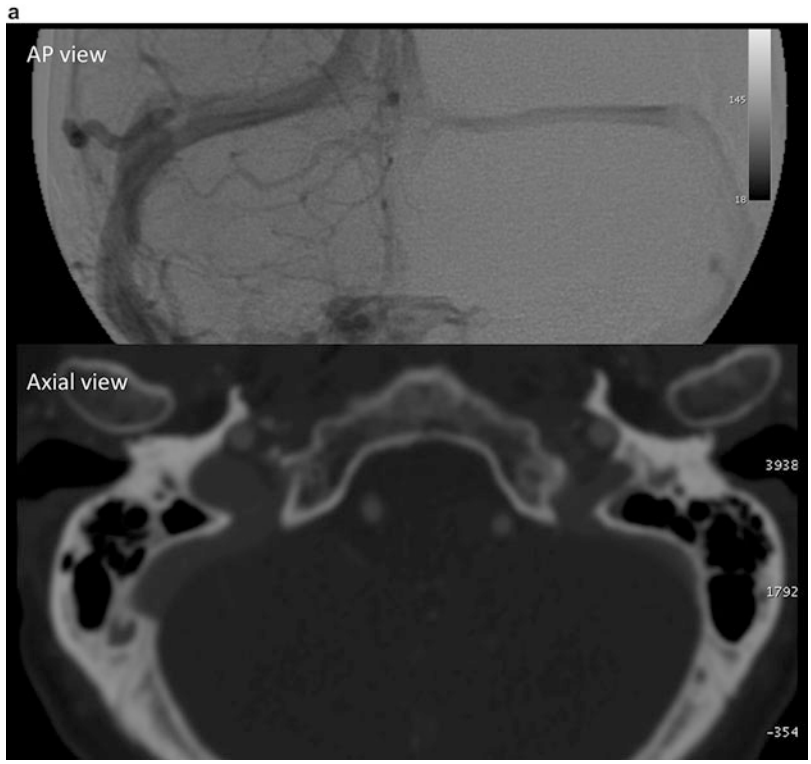


Fig. 15 (continued)

such cases the basal vein of Rosenthal drains into the lateral sinus or torcula (through tentorial sinus) or through the latero-mesencephalic vein.

The vein of Galen is a middle line single vein that receives the blood from the internal cerebral veins at the level of the quadrigeminal cistern. It can also receive the blood from the basal vein of Rosenthal directly. During its course it collects the blood from inferior pericallosal veins, medial occipital veins and from precentral veins and superior vermian veins (Duvernoy 1975; Lasjaunias et al. 2001; Rhoton 2000a, 2002b).

Intracranial Venous Variability and Anomalies

The intracranial venous system has a wide anatomic variability, rich anastomosis network, bidirectional flow, and capacity to adapt to flow changes. This makes the outcome of venous thrombosis unpredictable.

The post birth maturation has been discussed previously and is extremely important for the evaluation of the pediatric vascular system.

Variability is the rule for venous anatomy. The size, number and disposition of the intracranial veins are extremely variable among

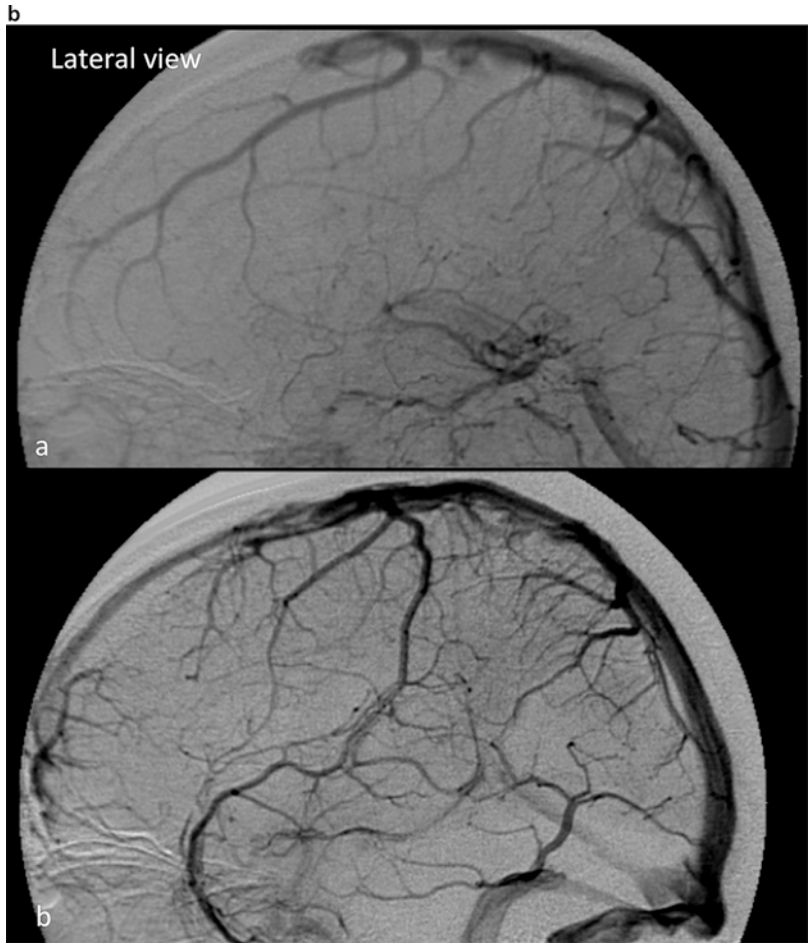


Fig. 15 (continued)

individuals and not predictable, which may explain different clinical outcomes in intracranial venous occlusion or in arteriovenous fistulae/malformations.

Hypoplasia or partial agenesis of the dural sinuses is frequent. The asymmetry of the transverse/sigmoid sinuses is a common example, and the development cause of this finding can be easily deducted from the size of the corresponding jugular foramen, differentiating it from acquired conditions (Fig. 15a).

The anterior third of the superior sagittal sinus is also variable and agenesis is associated with hypertrophy of frontal cortical veins that collect the venous blood from that area (Fig. 15b).

The cortical veins distribution is another example of extreme variability. For the cerebral convexity there may be a pattern of equal dominance of cortical veins, or of Trolard, Sylvian, Labbé dominance with a large vein draining into the superior sagittal sinus, cavernous sinus, or transverse sinus respectively (Fig. 15c).

Other venous variants include the persistence of prominent embryonic veins/sinuses, such as (Fig. 16a) (Lasjaunias et al. 2001, 2006; Raybaud 2010):

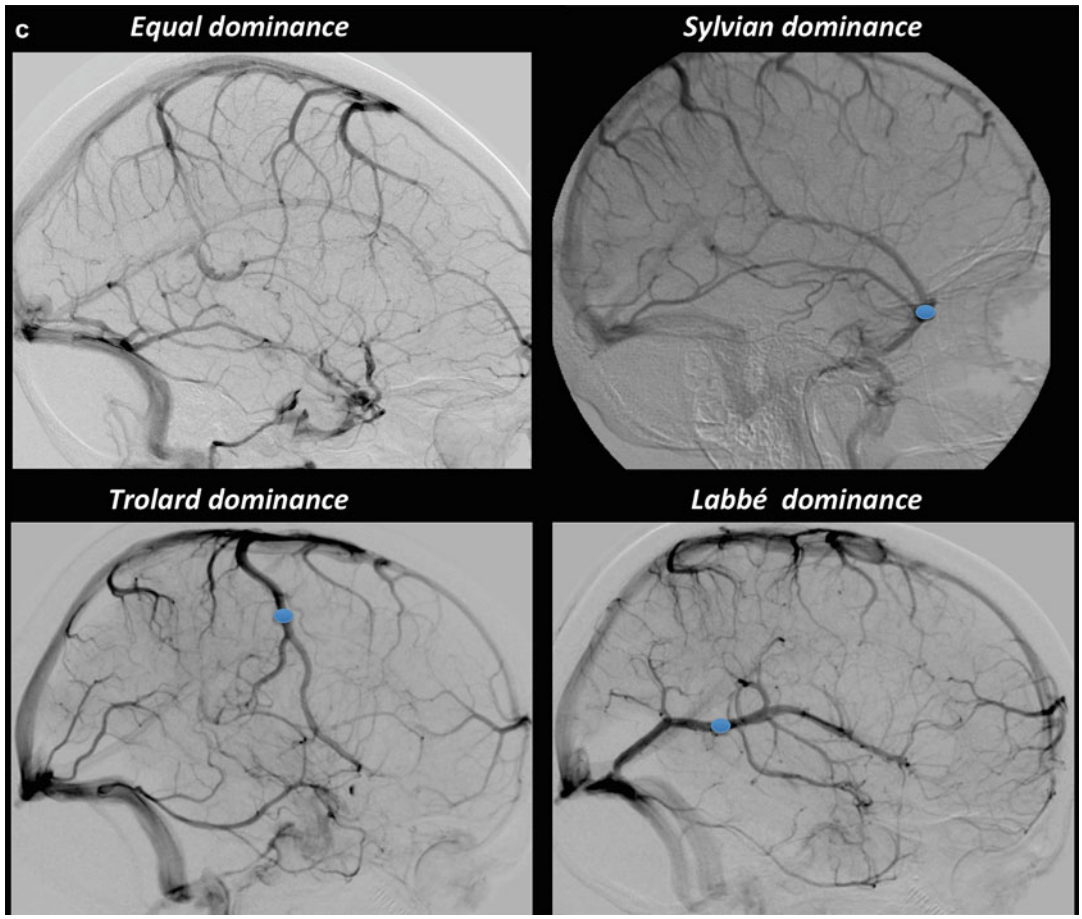


Fig. 15 (**a**) DSA venous phase antero-posterior view DSA and axial CT demonstrate (congenital) left transverse and sigmoid sinuses hypoplasia. (**b**) Venous variants. DSA venous phase lateral view shows a superior sagittal sinus partial agenesis (upper image) in comparison to a complete superior sagittal sinus (lower image). (**c**) Venous variants. DSA venous phase lateral view illustrates different patterns of the cerebrum convexity venous drainage (co-dominance and dominance)

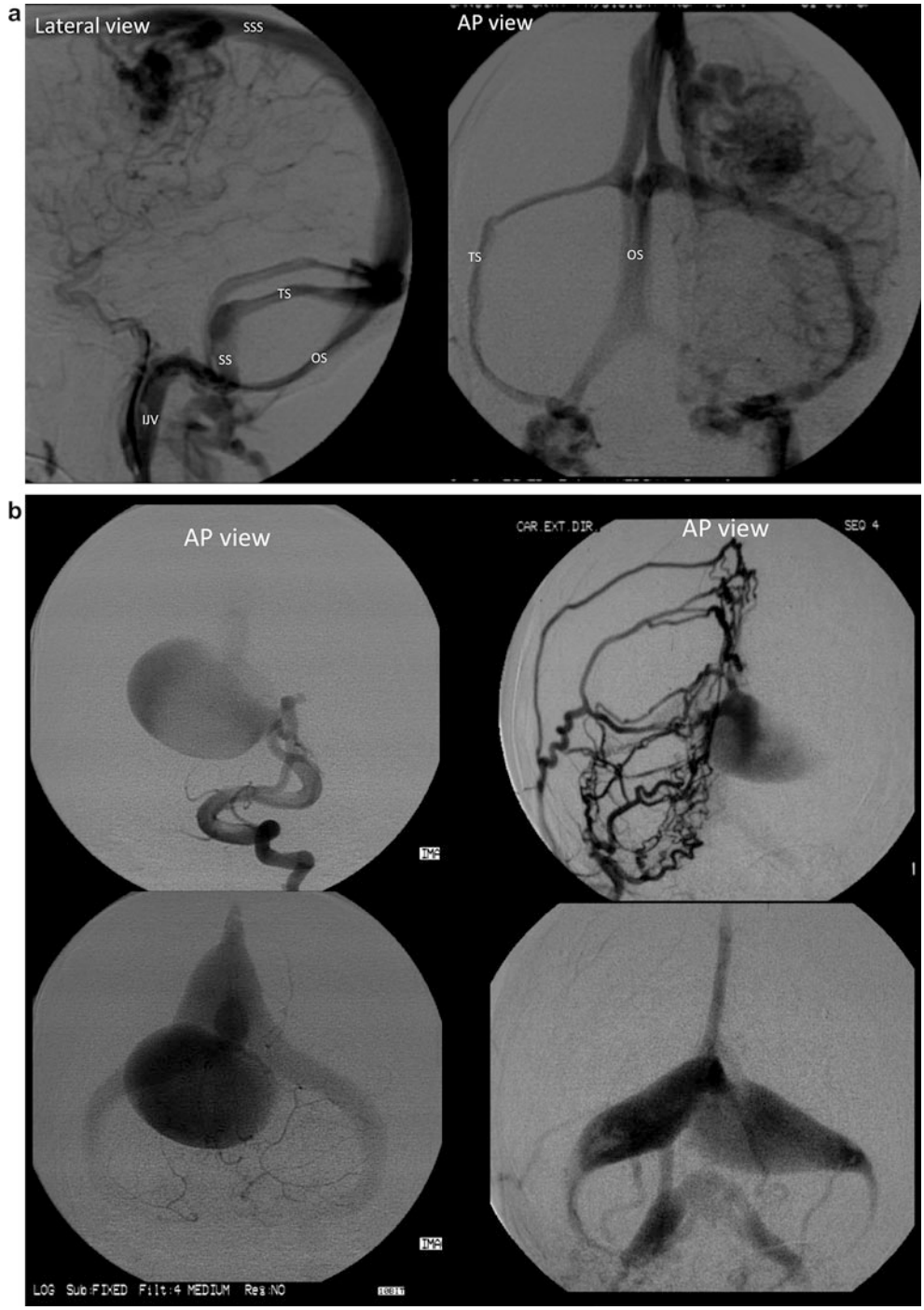


Fig. 16 (**a**) DSA venous phase lateral and anteroposterior view illustrating the persistence of a major occipital sinus. (**b**) DSA venous phase anteroposterior view showing the persistence of the Markowski's vein in association with VGAM (left images) and "balloon-shaped" dural – transverse sinuses and torcula – sinuses in association with dural sinus malformation with AV shunts (right images)

- Occipital sinus: Beginning at the torcula and flowing into the marginal or sigmoid sinus with communications to the internal jugular vein, clival plexus, and vertebral–suboccipital plexus
- Marginal "sinus": Which is a venous plexus around the foramen magnum, connected with the basilar plexus, sigmoid sinus, and occipital sinus
- Falcine "sinus": Which is a venous plexus sited within the falx cerebri and communicating the vein of Galen and the superior sagittal sinus
- Tentorial sinus: Which are transient structures, located within the tentorium draining the temporal lobe and diencephalon towards the torcula/transverse sinus and that regress after the development of the basal vein of Rosenthal

The persistence of embryonic veins/dural sinuses may occur as an isolated feature or in association with vascular malformations, such as the Vein of Galen aneurysmal malformation with the persistence of the Markowski's vein, or the dural sinus malformation with AV shunts with the "ballooning" appearance of the dural sinus representing a "frozen" anatomy at the time of the malformation development.

Other more complex venous anomalies include the Development Venous Anomalies (DVA) and the *sinus pericranii* (see ▶ Chap. 10, "Intracranial Vascular Malformations").

DVAs are extreme variations of the transmedullary venous development. They are common findings with an incidence of 2.5–3% in autopsy. The etiology is not completely understood, being accepted that represent an in uterus failure of normal venous development and/or venous thrombosis (Fig. 17) (Lasjaunias et al. 2001, 2006; Raybaud 2010).

They may be categorized into:

- Superficial DVAs accounting for ~70% of all DVA cases and drain the deep medullary regions into cortical veins.
- Deep DVAs corresponding to ~20% of all DVA cases and drain the subcortical medullary regions into deep venous collectors.
- Mixed (deep & superficial) DVAs accounting for up to 10% of cases. They are composed of a *caput medusae*, corresponding to a group of radial arranged dilated medullary veins converging into a single (or multiple) transmedullary venous collector.

They are characterized by the lack of normal surrounding venous system, they have intermingled normal brain tissue and they participate in the normal brain tissue venous drainage. They have been described in association with cavernomas, but also with Brain AVMs, cortical malformations and in syndromatic forms such as Sturge Weber syndrome and Blue Rubber Bleb Nevus (Lasjaunias et al. 2001, 2006; Raybaud 2010).

During the neonatal period they have a dynamic behaviour with flow changes, including increase in flow or disappearance inducing adjacent parenchymal changes, including the appearance of new changes or the regression of previous brain abnormalities. In adults, they are generally stable and asymptomatic. However, as any other intracranial venous structure, DVA may become symptomatic due to mechanical compression of neural structures causing hydrocephalus or cranial nerve palsies or due to flow changes with reduced flow associated with venous stenosis/thrombosis (which may cause any of the venous thrombosis complications described in the Venous Thrombosis chapter) or with increased flow whenever associated with an AV shunt.

The *sinus pericranii* (SP) are defined as large venous connection between the intra and extracranial venous drainage, formed by a network of thin-walled veins that form a varix on the external table of the skull, in which the blood may have a bidirectional flow (Fig. 18). Sinus pericranii are generally unique, asymptomatic, located at the midline at the frontal area. Multiple sinus pericranii are very rare (Lasjaunias et al. 2001, 2006; Raybaud 2010).

They may be categorized into two major groups:

- Accessory – SP with a small amount of venous flow crossing through
- Dominant – SP with a mainstream of intracranial venous blood crossing, playing a major role in brain venous drainage

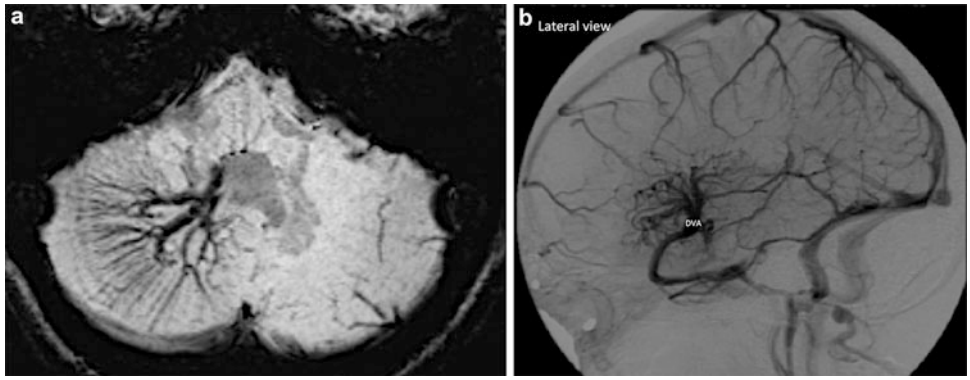


Fig. 17 (**a**, **b**) Developmental venous anomaly (DVA). (**a**) Axial SWI MR image showing a large cerebellar DVA. (**b**) DSA venous phase lateral view demonstrating a superficial DVA draining into the sylvian group (sylvian veins-cavernous plexus)

The diagnosis of *sinus pericranii* is clinical. Imaging may be useful to evaluate the flow with Doppler US, the bone defect with computed tomography (CT), the complete venous pattern with computed tomography venogram (CTV) or magnetic resonance venogram (MRV), or to look for other venous and brain anomalies with MRI. Digital subtraction angiography is generally not needed unless treatment is considered and precise evaluation of the sinus pericranii role for the brain drainage is warranted (Lasjaunias et al. 2001, 2006).

Collateral Circulation

The collateral circulation is composed of vascular redundancies from pre-existing arterial and venous vessels, with bidirectional flow (depending on the pressure gradients), allowing alternative vascular pathways maintain the blood flow, overcoming vascular stenosis/occlusions. Each individual has a singular arterial and venous collateral status.

The arterial collateral development (collaterogenesis) occurs at a very early stage, around the 13–14 gestation weeks. This process is genetic mediated, being chromosome 7 and the Dce1 locus important regulators of collateral arteries. Several acquired conditions, such as aging, chronic hypertension, metabolic syndrome and diabetes, alter the collateral status of individuals by decreasing of collateral number and/or diameter.

The most common arterial anastomotic pathways include external–internal carotid (or ophthalmic) arteries anastomoses, external carotid–vertebral artery anastomoses, the persistence of embryonic internal carotid artery–vertebro-basilar anastomoses (trigeminal, hypoglossal, proatlantal type 1 and 2 arteries), the circle of Willis, and leptomeningeal anastomosis through cortical/pial or deep arteries (Fig. 18).

The collateral venous status is even more unpredictable than the arterial. The venous anatomy is highly variable among individuals, specially the superficial venous system. The venous system is highly prone for anastomoses allowing multidirectional flow accordingly to local flow needs. The major anastomotic venous pathways are extracranial–intracranial anastomoses, through cavernous plexuses, meningeal, diploic, and emissary veins of the skull; superficial–deep venous system anastomoses through transmedullary veins; infratentorial–supratentorial venous systems, through large anastomotic veins such as lateral mesencephalic vein and basal vein of Rosenthal; infratentorial–spine venous system, through medullary and spinal veins. There are also anastomoses inside each venous system, such those between veins of the deep venous system, through venous circle of Willis, or through the peripheral venous network surrounding the brain stem, and between cortical veins through pial anastomoses (Figs. 19 and 20).

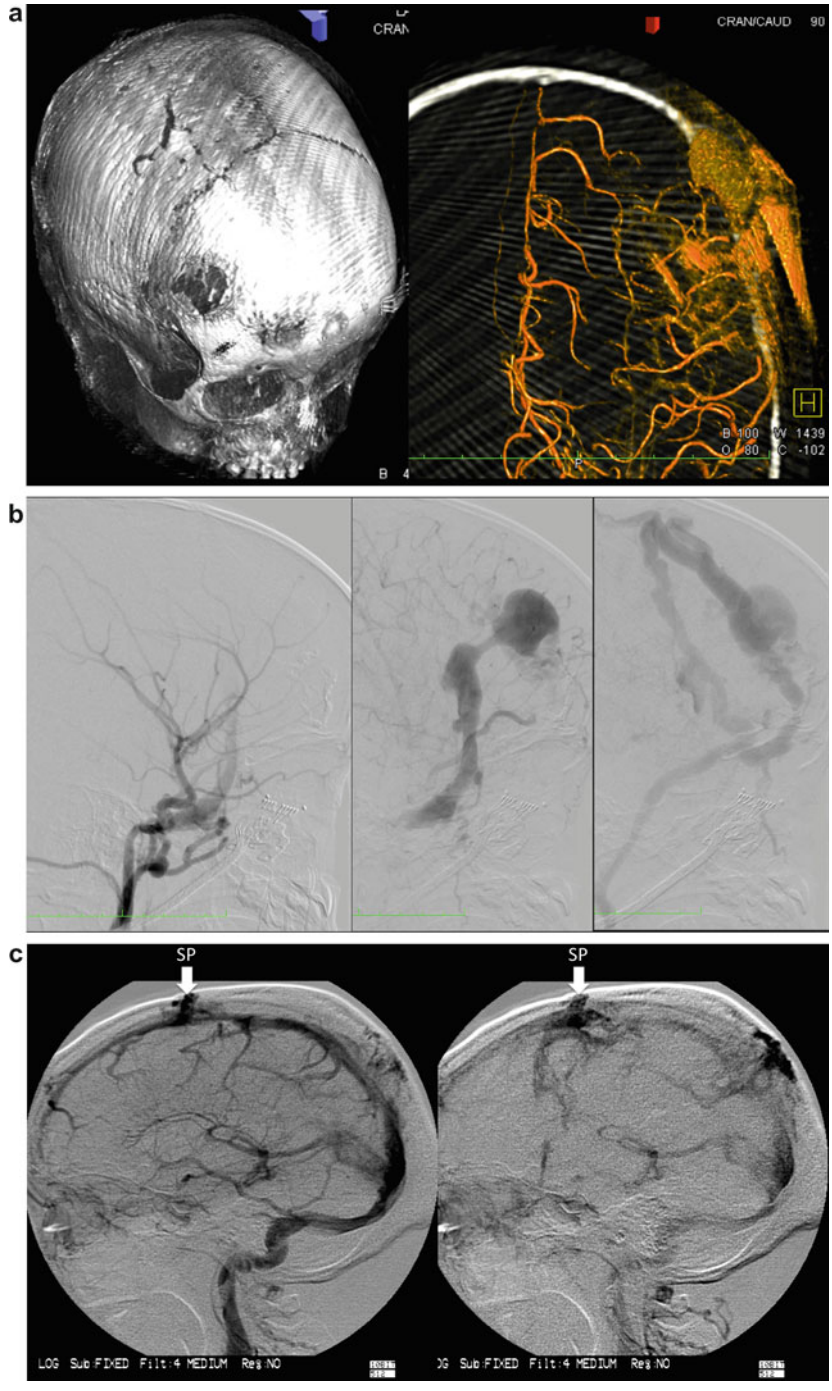


Fig. 18 (**a**, **b**) Sinus pericranii (SP). (**a, b**) DSA of left frontal SP associated with a congenital middle fossa AVF. (**a**) The 3D bone reconstruction shows the bone defect, and the axial images demonstrate the venous pouch crossing the bone defect. (**b**) Lateral view of common carotid artery injection demonstrating the presence of a high flow middle fossa AVF fed by ECA branches and draining thought a enlarge convexity vein toward the bone defect and into the extracranial scalp veins. (**c**) DSA Lateral view of ICA injection showing a small SP at the cranial vault communicating the superior sagittal sinus with the extracranial venous system. There are also multiple enlarged transosseous veins at the posterior third of the superior sagittal sinus communicating with the extracranial venous system

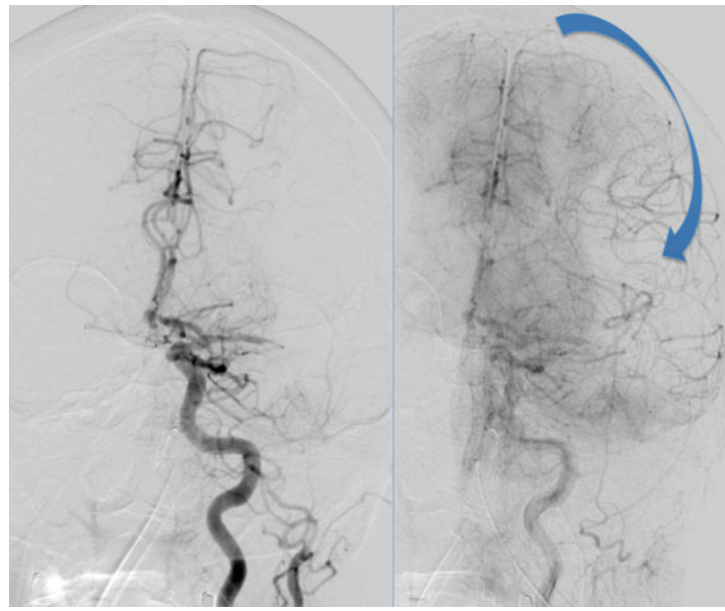
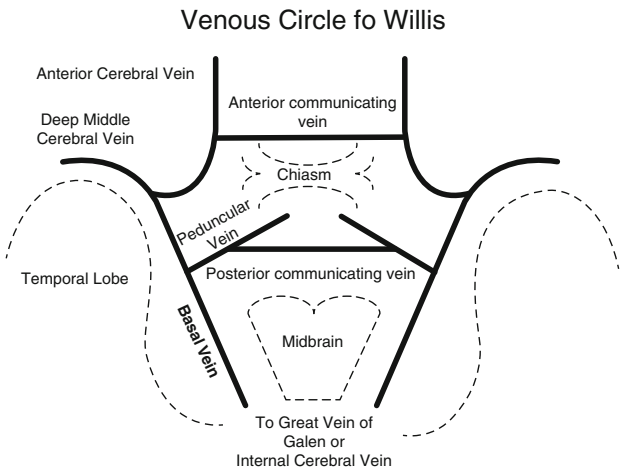


Fig. 19 Arterial collateral circulation. DSA anteroposterior view after the left ICA contrast injection shows a distal M1 occlusion in the setting of acute ischemic stroke and the retrograde filling of distal MCA branches through leptomeningeal anastomosis with ACA



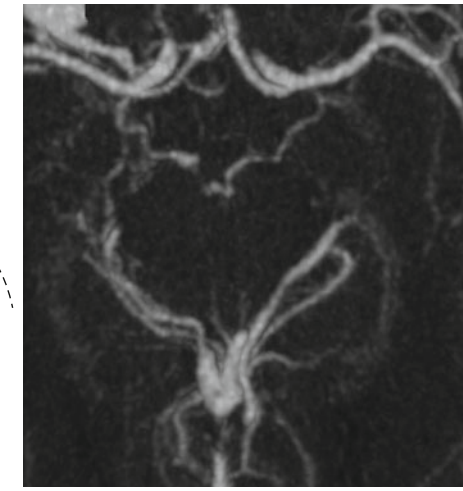


Fig. 20 Venous collateral circulation. Schematic representation (left image) of the venous "circle of Willis" and the corresponding finding in 3D MIP axial images obtained with 3D DSA (right image). (Courtesy of Dr.ª Teresa Nunes – Portugal)

References

Duvernoy HM. Human brain stem vessels. 2nd edition. Berlin: Springer 2010.

Duvernoy HM. The superficial veins of the human brain: veins of the brain stem and of the base of the brain. Berlin: Springer; 1975.

Lasjaunias P, Berenstein A, ter Brugge KG. Surgical neuroangiography. 1. Clinical vascular anatomy and variations. 2nd ed. Berlin: Springer; 2001.

Lasjaunias P, Berenstein A, ter Brugge KG. Surgical neuroangiography. 3. Clinical and interventional aspects in children. 2nd ed. Berlin: Springer; 2006.

Milner R, editor. Cerebral angiogenesis: methods and protocols, methods in molecular biology, vol. 1135. New York: Springer Science+Business Media; 2014.

Raybaud C. Normal and abnormal embryology and development of the intracranial vascular system. Neurosurg Clin N Am. 2010;21(3):399–426.

Rhoton AL Jr. The cerebellar arteries. Neurosurgery. 2000a;47(3 Suppl):S29–68.

Rhoton AL Jr. The posterior fossa veins. Neurosurgery. 2000b;47(3 Suppl):S69–92.

Rhoton AL Jr. The supratentorial arteries. Neurosurgery. 2002a;51(4 Suppl):S53–120. Review

Rhoton AL Jr. The cerebral veins. Neurosurgery. 2002b;51 (4 Suppl):S159–205. Review

Rhoton AL Jr. The cavernous sinus, the cavernous venous plexus, and the carotid collar. Neurosurgery. 2002c;51 (4 Suppl):S375–410

Ribatti D, editor. Vascular morphogenesis: methods and protocols, methods in molecular biology, vol. 1214. New York: Springer Science+Business Media; 2015.