Les artères de la moelle épinière cervicale et du tronc cérébral. Anatomie angiographique et de développement

Siclari, Francesca

Section de Médecine Fondamentale
Département de Physiologie
cellulaire et de Métabolisme
Division d'Anatomie

Thèse préparée sous la direction du Professeur
Jean Henri Dominique Fasel

"LES ARTERES DE LA MOELLE EPINIERE CERVICALE ET DU TRONC CEREBRAL. ANATOMIE ANGIOGRAPHIQUE ET DE DEVELOPPEMENT."

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Francesca SICLARI

Lugano (TI)

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- Burger I, Siclari F, Gregg L, Gailloud P. Bilateral segmental agenesis of the vertebrobasilar junction: developmental and angiographic anatomy. Am J Neuroradiol. 2007;28(10):2017-22.

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CONTENTS

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RESUME

Introduction

La vascularisation artérielle de la moelle cervicale et du système vertébro-basilaire a été étudiée de façon extensive au cours des siècles passés et apparaît comme une entité bien établie, peu sujette à de nouvelles interprétations. Parmi les travaux les plus fréquemment cités à ce sujet figurent ceux d'Adamkiewicz (1, 2) et de Kadyi (3), parus à la fin du 19ème siècle, avec un accent particulier mis sur la variabilité des contributions radiculaires à la vascularisation médullaire. L'embryologie des vaisseaux du système nerveux central fut étudiée de façon détaillée par Padget (4) et Congdon (5) sur l' embryon humain, et par Moffat (6, 7) sur le rat.

De nos jours, les techniques d'imagerie vasculaire permettent une excellente visualisation de l'ensemble de l'arbre artériel. Le radiologue se trouve ainsi fréquemment confronté à des variations artérielles complexes, dont l'origine est peu claire, et qui sont parfois difficiles à distinguer de processus pathologiques. Une connaissance approfondie de l'embryologie vasculaire est essentielle afin de comprendre l'origine de ces variations et de surmonter ces difficultés diagnostiques. En se basant sur les travaux de Kadyi (3), Padget (4) et Moffat (6, 7), le présent travail élabore plusieurs concepts embryologiques originaux, et les applique à des variations artérielles du système vertébro-basilaire observées en pratique clinique.

Le texte de cette thèse se divise en trois parties. La première partie présente une revue de l'anatomie artérielle de la moelle cervicale et du tronc cérébral. La deuxième partie retrace le développement embryonnaire des vaisseaux médullaires et du système vertébro-basilaire. Dans la troisième partie, trois types de variations artérielles du système vertébro-basilaire sont

décrits et illustrés par des images angiographiques. La discussion porte sur leurs mécanismes de formation.

Matériel et méthodes

Revue de la littérature

Nous avons considéré les travaux originaux relatifs à l'anatomie et à l'embryologie vasculaire de la fosse postérieure et de la moelle cervicale, ainsi que les travaux contenant des descriptions de variations artérielles dans ce territoire. La recherche a été effectuée d'une part dans la banque de données « Medline » (1948 à 2008). D'autre part, les travaux anatomiques originaux ayant été publié en grande partie avant 1948, nous avons étendu la recherche à la bibliographie personnelle des directeurs de thèse et à celle de la bibliothèque de la faculté de médecine de l'Université de Genève et de l'Université de Johns Hopkins à Baltimore, Etats-Unis. D'autres publications ont été inclues à partir de la liste de références contenue dans ces ouvrages. En nous basant sur cette littérature, nous avons réalisé les deux premières parties de cette thèse, qui constituent une revue de l'anatomie et l'embryologie vasculaire du tronc cérébral et de la moelle cervicale.

Identification des variations artérielles

Nous avons ensuite identifié, parmi les angiographies effectués dans la division de neuroradiologie de l'Université de Johns Hopkins à Baltimore (Etats-Unis), des variations artérielles qui résultent des étapes embryologiques mentionnées dans la première et deuxième partie.

Résultats

Trois types de variations artérielles, illustrées par 21 cas cliniques, ont été identifiées et mises en relation avec des concepts embryologiques.

L'agénésie bilatérale de l'artère vertébrale distale

L'agénésie bilatérale de l'artère vertébrale distale est une variation exceptionnelle. Nous en illustrons deux exemples, caractérisés par l'absence complète non seulement des segments V4, mais également de l'artère basilaire proximale et de sa connexion avec l'artère cérébrale postérieure. L'origine de cette variation devient compréhensible lorsqu'on admet que l'artère vertébrale distale est la branche radiculaire antérieure de C1, et que l'artère basilaire proximale résulte de la fusion des deux rameaux ascendants de cette branche radiculaire antérieure. Ce concept fut établi par Kadyi en 1889 (3). Nous suggérons que l'absence bilatérale de l'artère vertébrale distale résulte de l'agénésie de la branche radiculaire antérieure de C1. De même, nous nous référons aux travaux de Moffat (6) sur le développement de l'artère cérébrale postérieure, pour montrer qu'il s'agit dans ces deux cas de variations d'origine congénitale, reflétant des contraintes hémodynamiques particulières durant la période embryonnaire. Afin d'illustrer la nature congénitale de ces deux variations, nous fournissons un exemple d'absence bilatérale acquise du segment V4, et montrons comment distinguer les deux cas de figure.

Les variations des artères spinales latérale et postérieure en relation avec l'anatomie de développement de l'artère vertébrale distale

Les origines basses de la PICA, au niveau C1 et C2, ainsi que les duplications et certains trajets aberrants de l'artère vertébrale sont classiquement attribués aux variations de l'artère spinale latérale (8). L'artère vertébrale distale et sa branche principale, la PICA, dérivent

embryologiquement de la branche spinale de l'artère proatlantale. Cette dernière est un vaisseau embryonnaire, dont le trajet est parallèle à celui du premier nerf cervical, et qui assure de façon transitoire l'apport sanguin à la circulation postérieure. Comme évoqué par Kadyi (3), l'artère spinale postérieure correspond à la branche descendante de l'artère radiculaire postérieure du premier nerf cervical. En nous basant sur ce concept, nous postulons que les variantes susmentionnées peuvent résulter non seulement des variations de l'artère spinale latérale, mais également de variations de l'artère spinale postérieure, et que les deux types de variantes ont un aspect angiographique distinct.

Les variations en relation avec l'anastomose primitive latérale vertébro-basilaire de Padget

Comme son nom l'indique, l'anastomose primitive latérale vertébro-basilaire de Padget (PLBA) est une structure embryonnaire, décrite par Padget en 1948 (4), qui constitue une anastomose longitudinale entre les artères vertébrale et basilaire primitives. Dorcas Hager Padget basa ses recherches sur 17 embryons humains étudiés à différents stades de développement. Utilisant une méthode de reconstruction graphique, elle fut capable de reconstituer pour la première fois le développement des vaisseaux cérébraux de façon détaillée. Elle postula que la persistance partielle de la PLBA pouvait expliquer certaines variations du système vertébro-basilaire. Plus tard, Moffat reconnût que la PLBA et l'artère spinale latérale constituaient les portions intra- et extracraniennes respectives du même axe vasculaire (6). Dans cette dernière partie, nous décrivons et illustrons cinq variations du système vertébro-basilaire, comprenant des variantes d'artère trigéminée persistante, des variations d'origine des artères cérébelleuses ainsi qu'une duplication de l'artère basilaire proximale. Nous proposons que ces variations résultent de la persistance de la PLBA, comme anticipé par Padget (4).

Conclusions

La présente thèse montre qu'une connaissance approfondie de l'anatomie de développement des vaisseaux cérébraux permet de comprendre des variations artérielles rencontrées en pratique clinique, et de les distinguer des variantes acquises (pathologiques). Ceci a des implications pratiques pour les professionnels dans les domaines de la neuroradiologie, de la neurochirurgie et de la neurologie.

PART I: NORMAL ANATOMY

THE ARTERIAL VASCULARIZATION OF THE CERVICAL SPINAL CORD AND BRAIN STEM

The following section outlines the normal arterial vascularization of the brainstem and spinal cord. Each major artery is described separately. Only the most relevant variations are mentioned in this chapter.

The arterial vascularization of the cervical spinal cord

The blood supply of the spinal cord can schematically be divided into three longitudinal vascular axes, which comprise the *anterior spinal artery* (ASA) and the two *posterior spinal arteries* (PSA). At the upper cervical level, an additional paired longitudinal artery termed *lateral spinal artery* (LSA) exists. These longitudinal axes are supplied by segmental vessels at various levels of the spinal cord.

The longitudinal arterial axes

The anterior spinal artery

The ASA results from the junction of two small anterior spinal rami originating from the intracranial portion of each vertebral artery (VA). These rami course ventromedially to form a single descending channel that runs along the ventral sulcus of the spinal cord. In practice, there is often, if not always, an asymmetry in caliber of these two rami, and it is not unusual to see the ASA as the continuation of a single VA branch. The newly formed vessel runs in the subpial space of the ventral sulcus, dorsal to the anterior spinal veins, and has a fairly straight course, although deflections towards the junction of the segmental feeding vessels

may occur (9, 10). Its diameter generally gradually tapers down at the upper thoracic level (10-12), but increases in size immediately below its junction with anterior radiculomedullary arteries (9, 10, 13). Although classically described as a continuous system, the ASA is typically discontinuous in some portions of its course, particularly at the thoracic level (10, 14-16).

The posterior spinal artery

The two PSAs, which are about the third the size of the ASA (10, 17), generally originate from the inferior or posterior aspect of the VA, near but proximal to its point of dural penetration, or alternatively from the posterior inferior cerebellar artery (PICA). When the PSA originates from the VA, the PSA and the VA take a parallel ascending course until they reach the lateral surface of the medulla oblongata. The VA then passes over the anterior aspect of the pontomedullary junction, while the PSA turns posterolaterally and, after a sharp curve, courses caudally along the dorsal aspect of the spinal cord. At the apex of the curve, the PSA generally gives off an ascending branch that anastomoses with the PICA at the level of the restiform body (18). The paired PSAs thus appear as two parallel anastomotic chains on the posterior-lateral aspect of the spinal cord, close and medial to the attachment of the posterior nerve roots.

The lateral spinal artery

The LSA was first differentiated from the PSA by Kadyi in 1889 (3), and later given the name of LSA. It lies on the lateral aspect of the spinal cord, ventral to the posterior cervical nerve roots, and parallel to the spinal component of the eleventh cranial nerve. The LSA has usually several lateral segmental connections with the VA at C2, C3, and/or C4. Cranially, it ends into the intradural portion of the VA at C1, and/or joins the PICA. Caudally, at the level of C4 or C5, it turns dorsally to pass behind the spinal nerve roots and join the ipsilateral PSA.

A number of variations at the vertebrobasilar junction, such as VA duplications, intradural courses of the distal VA, as well as low or duplicated PICA origins, have traditionally been attributed to variant courses of the LSA (8).

The segmental vascularization

General organization

As outlined above, the longitudinal spinal axes are supplied by segmental vessels at various spinal levels. At the cervical level, the main contributors originate from the VA, the ascending cervical arteries, and the deep cervical arteries. Typically, these vessels give off a branch that enters the intervertebral (or neural) foramen and then divides into a vertebral branch supplying the vertebral body and a radicular branch that pierces the dura and accompanies the spinal nerve root. The radicular branch further divides into anterior and posterior radicular branches, coursing along the anterior and posterior nerve roots, respectively. At the adult stage, only a few of these segmental vessels provide a significant contribution to the anterior and posterior spinal axes. When they do so, they are generally referred to as *radiculomedullary arteries* in order to distinguish them from radicular arteries that merely supply the nerve roots and do not contribute to the longitudinal spinal axes, and from radiculomeningeal arteries that supply the nerve root and the spinal meninges (Fig.1).

Radiculomedullary arteries are usually asymmetric and reach the cord from either the left or the right side, but generally not from both sides at the same segmental level. In those rare instances in which arteries from both sides join the ASA at the same segment, a diamond-shaped pattern may occur (9, 19). This pattern is most often observed angiographically in

children. Similarly, it is either the anterior or the posterior vessel that reaches the cord but generally not both vessels at the same level (3, 17). When anterior and posterior spinal contributors are provided by the same radicular artery, the variant takes the name of an artery of Lazorthes. The course of radicular branches at the cervical level is relatively horizontal while their trajectory becomes progressively steeper as one progresses in a caudal direction. At the spinal cord level, they divide into a prominent descending branch and a smaller ascending branch that both join the longitudinal axis. This results in a prominent cranial loop with the typical “hairpin” appearance on angiograms (Fig. 2). The cranial loop has a wider angle in anterior radiculomedullary arteries than in the posterior ones.

By analogy, at the thoracic and lumbar levels, the medullary blood supply comes from the lumbar and intercostal arteries, respectively. At the level of the vertebral body, these vessels give off a posterior branch termed the *dorsospinal artery*, before continuing as the intercostal or lumbar artery *per se*. The dorsospinal artery itself divides into a dorsal branch supplying the paravertebral musculature and a radicular branch entering the intervertebral foramen.

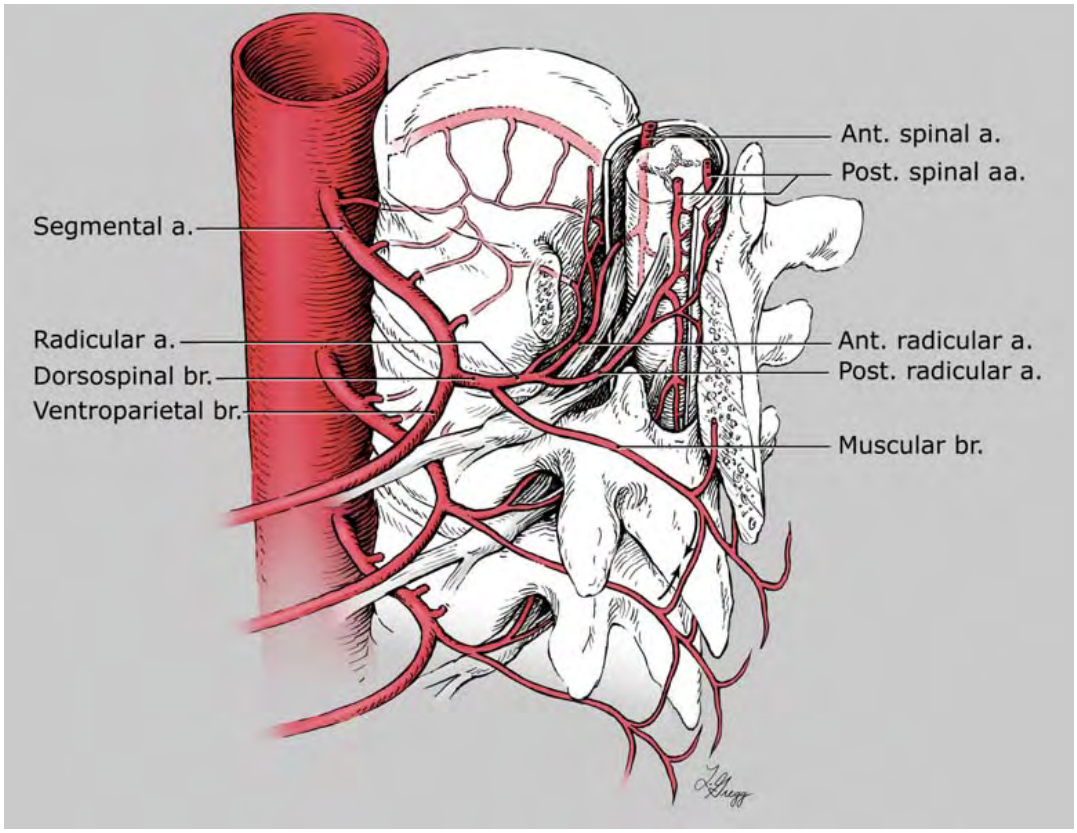


Fig. 1 the extrinsic vascularization the spinal cord

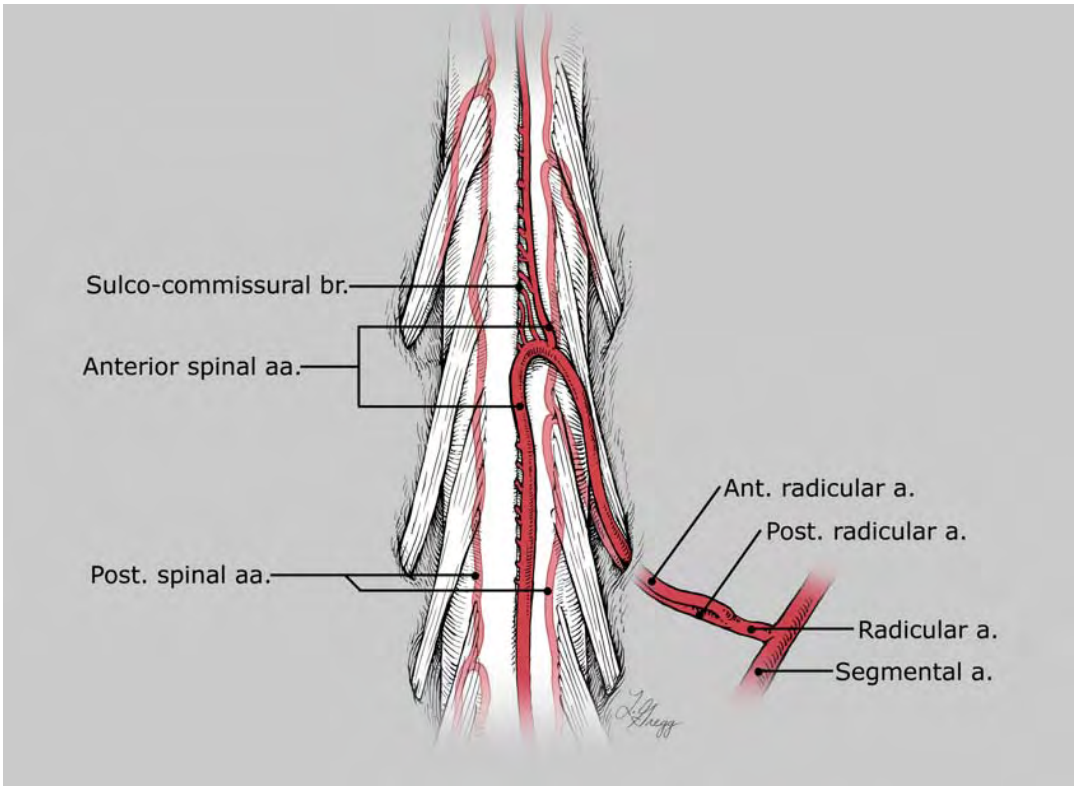


Fig. 2: branching pattern of the anterior radiculomedullary artery: the ascending branch is smaller, the descending branch displays the typical hairpin loop

Anterior radiculomedullary arteries

The number of anterior radiculomedullary arteries is extremely variable between individuals and at various spinal cord levels (Table 1). The upper cervical region (C1-C3) generally relies on blood supply from the two anterior spinal rami from the distal VA; anterior radiculomedullary arteries at this level are generally rare or absent. One can usually find 2 to 3 arteries at the C4 to C6 level that arise from the vertebral or ascending cervical artery. Below the C6 level, arteries contributing to the anterior spinal axis generally originate from the ascending cervical artery or the deep cervical artery. Lazorthes (20) described a large and particularly constant anterior radiculomedullary artery at the C6 level, which he called *the artery of the cervical enlargement* (12, 21, 22).

Author	**Year**	**Number**	**Level**
Adamkiewicz (2)	1882	3	C4, C5, C8
Kadyi (3)	1889	2	C4- C7, most constantly C5 or C6
Suh and Alexander (11)	1939	2 or 3	C3-C6
Zulch (23)	1954	1 or 2	C6, C7
Woollam and Millen (21)	1958	1, 3-5 minor	C6, C7 (major), above C6 (minor)
Lazorthes (20)	1957	1 to 3	C4-C8, most constantly C6
Gillilan (9)	1958	2	C3, C5-C6
Perese and Fracasso (12)	1959	1 to 5	C3-C7
Corbin (14)	1961	3 to 6	C4-C8
Jellinger (17)	1966	2 to 3	C5-C7
Chakravorty (10)	1971	2 or 3	C4-C6
Domisse (22)	1975	1 to 3	C4-C7

Table 1 (modified after Chakravorty (10)): number and most frequent origin of anterior radiculomedullary arteries at the cervical level

Posterior radiculomedullary arteries

Posterior radiculomedullary arteries are more numerous and have a smaller diameter than their anterior counterparts (3, 17, 24). Again, their number is variable (Table 2), but there is usually one major radicular artery at the C4 level, and one or two between the C2 and C6 levels.

Author	**Year**	**Number**	**Level**
Adamkiewicz (2)	1882	1 or 2	C6 or C7
Kadyi (3)	1889	1 or 2	C4-C7
Bolton (13)	1939	1	C4
Woollam and Millen (21)	1958	2 to 5	C2, C5-C7
Chakravorty (10)	1971	1 or 2	C2-C6, most constantly C4
Domisse (22)	1975	1 to 4	C5-C7

Table 2 (modified after Chakravorty (10)): number and most frequent origin of posterior radiculomedullary arteries at the cervical level

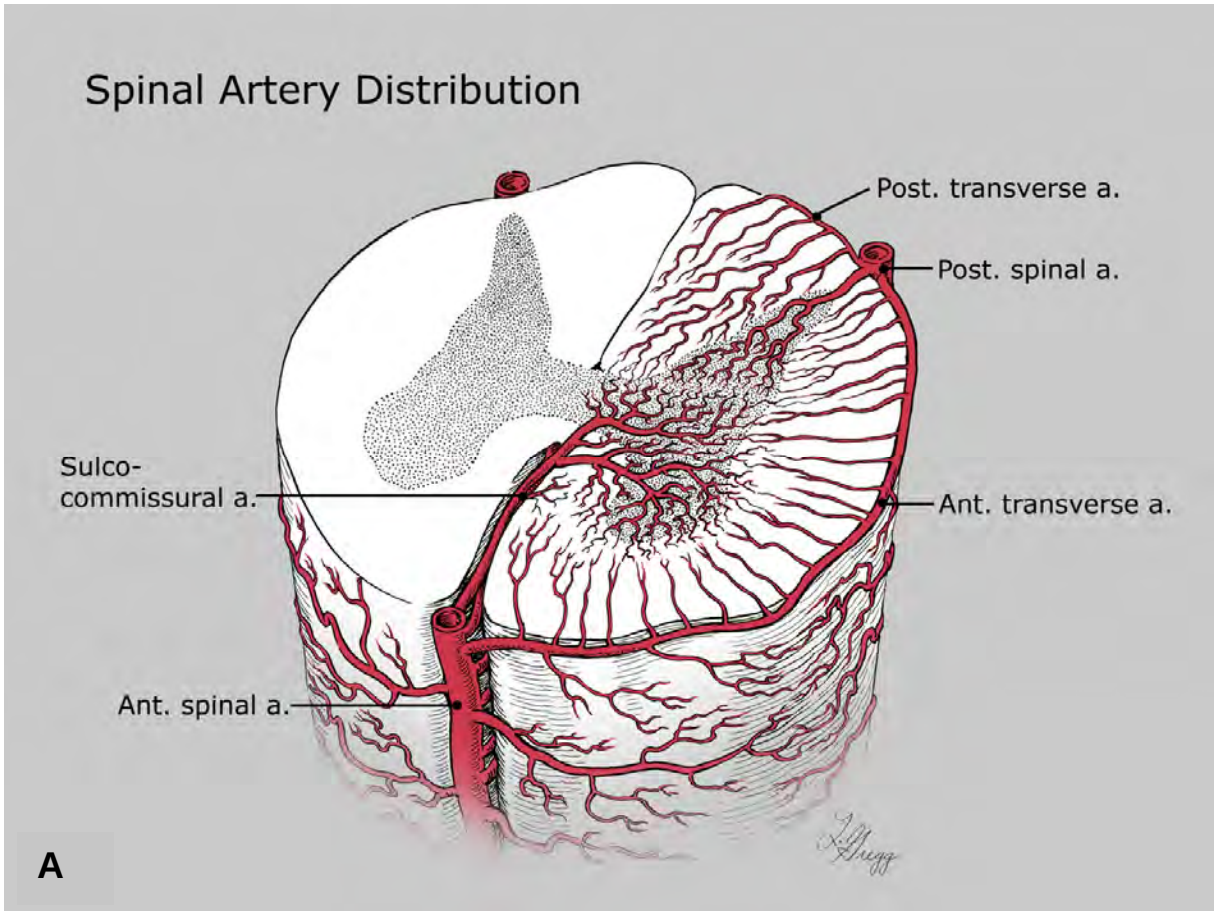
The intrinsic vascularization

The intrinsic vascularization of the spinal cord is derived from central arteries, also referred to as *central sulcal arteries* or *sulcocommissural arteries*, and peripheral arteries.

Central arteries

At each segmental level, the ASA gives off a central branch that aims posteriorly in the anterior median fissure to reach the anterior white commissure of the spinal cord, and then turns laterally to one side to enter the grey matter of the ventral horn (Fig. 3). These arteries

constitute the so called “centrifugal system of Adamkiewicz” (2), as they course from the centre towards the periphery. Arteries going to the right generally alternate irregularly with arteries going to the left. Sometimes, a common stem giving rise to a left and a right central artery can be observed, this most frequently occurs at the lumbar and sacral levels (14, 15). When the ASA is double, the central branches invariably go to the ipsilateral side (24). On reaching the grey matter, central arteries divide into an ascending and a descending branch that extend upwards and downwards so that there is considerable overlap at the capillary level (24). Central arteries are generally particularly large, numerous and densely packed in the regions of the cervical and lumbar enlargement of the cord (15, 17).



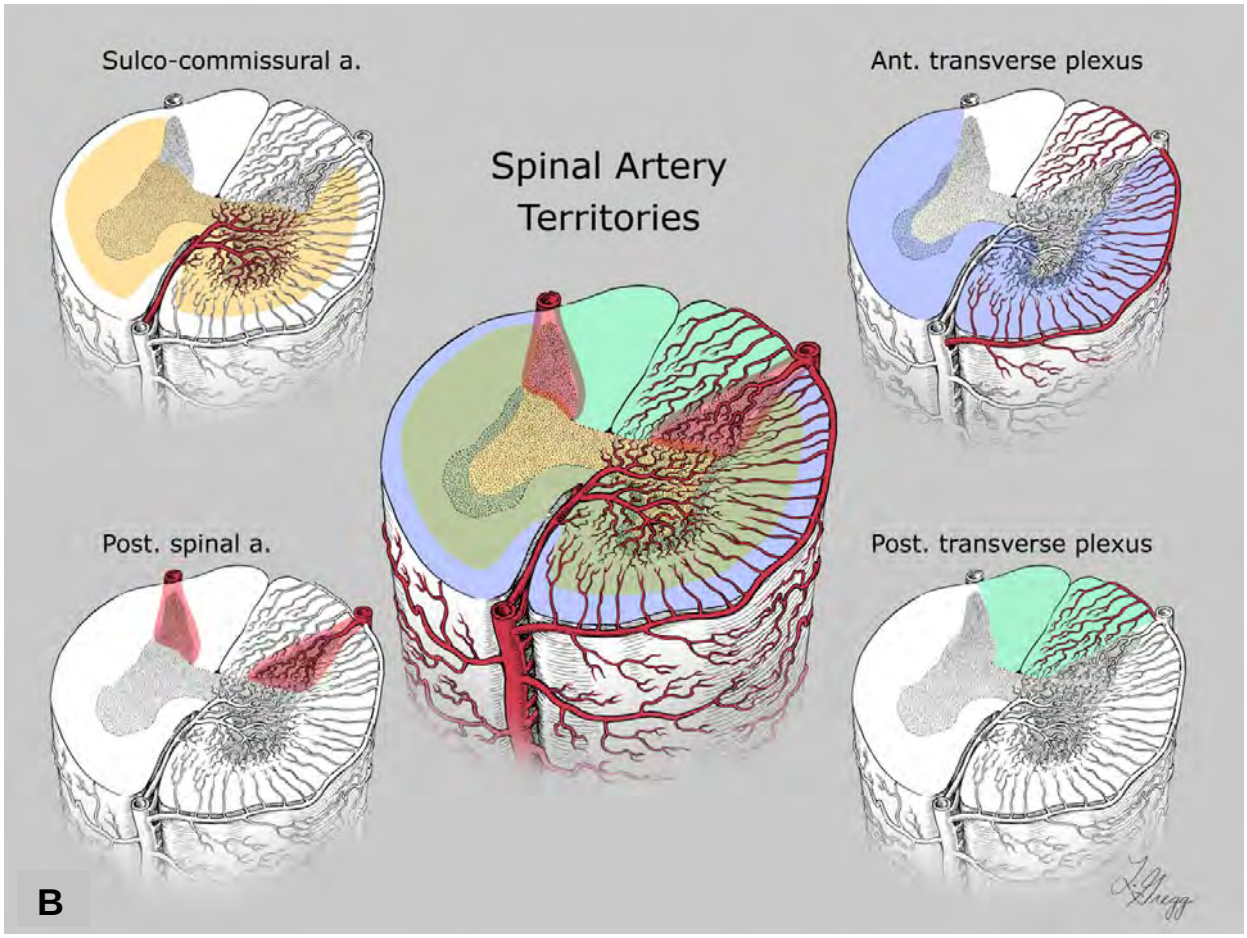


Fig. 3: spinal artery distribution

A, intrinsic blood supply of the spinal cord

B, vascular territories of spinal arteries

Peripheral arteries

The ASA and PSA each give off small lateral pial branches that run along the circumference of the cord and constitute the so called "*vasocorona"* or the "*centripetal system* of Adamkiewicz" (2). They give off perpendicular branches that penetrate the spinal cord at right angles and supply the outer rim of the spinal cord (9).

Anastomoses between central and peripheral arteries

The territories of the central arteries and peripheral arteries overlap in an area consisting of the inner portion of the white matter and the outer edge of the grey matter (except for the posterior halves of the post horns, which are supplied by peripheral arteries). The widest overlap exists in the posterior and lateral columns (19) (Fig. 3B).

The arterial vascularization of the brain stem

The vertebral artery

The VA most often originates from the subclavian artery, generally as its first branch. It is traditionally divided into four segments. The first segment (V1, extraosseous segment or *pars praevertebralis*) ascends superomedially from its origin at the subclavian artery, passes anterior to the transverse process of the C7 vertebra and usually enters the transverse foramen at the C6 level. The second segment (V2, foraminal segment or *pars transversaria*) follows a rather straight course through the transverse foramina from C6 to C3. The third segment (V3, atlanto-axial segment) starts above the transverse process of C3, where the artery proceeds laterally before entering the transverse foramen of the axis and atlas. The *pars atlantica* of the VA begins at its point of entry into the transverse foramen of the atlas. It courses posteromedially, and then turns upward, partly covered by the atlanto-occipital membrane, and penetrates the dura. The fourth segment (V4, intradural segment or *pars subarachnoidalis*) enters the posterior fossa through the lateral aspect of the foramen magnum. It then takes an anterior, superior and medial course and generally joins the contralateral VA at the level of the pontomedullary junction to form the basilar artery (Fig. 4). As outlined in the previous section, the VA supplies the anterior and posterior spinal axes via its radiculomedullary branches (Fig. 5). Additionally, the V2 and V3 segments give off

muscular branches that collateralize extensively with branches from the occipital, ascending pharyngeal, and deep cervical arteries, thus establishing anastomoses between the vertebral and external carotid circulation (25, 26).

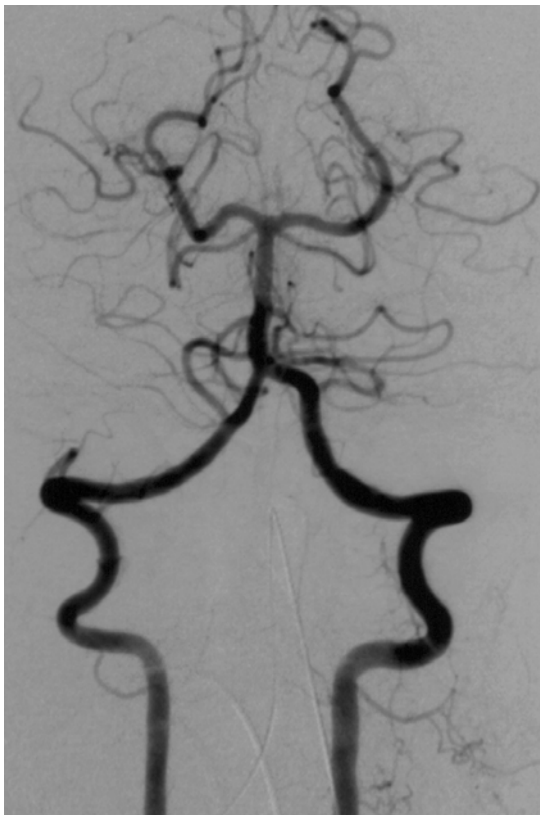


Fig. 4: DSA, vertebral injection, anteroposterior view, displaying the anatomy of the distal vertebral arteries

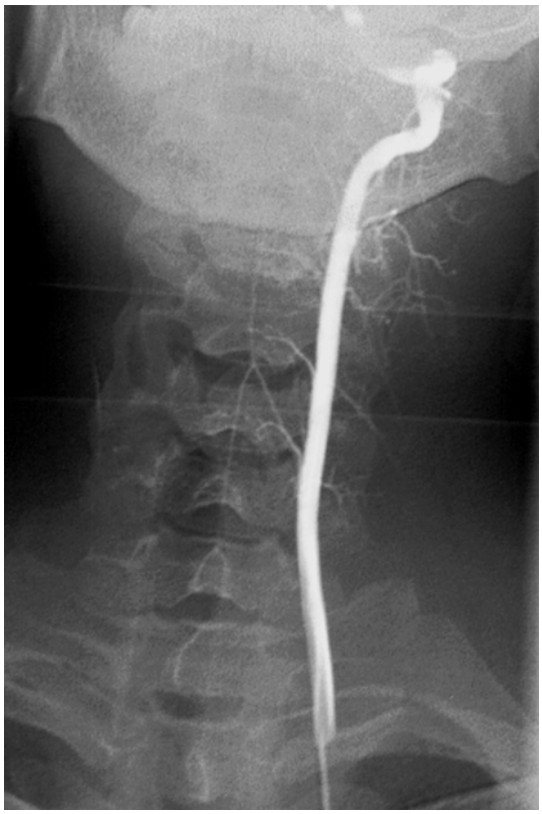


Fig. 5: DSA, left VA injection, nonsubtracted view, showing a prominent anterior radiculomedullary artery originating from the VA and feeding the anterior spinal axis (artery of the cervical enlargement)

Variations of course and origin

The right VA generally arises 25 mm distal to the branching point of the brachiocephalic trunk. Its origin is frequently shifted laterally or medially, each occurring in about 4% of cases (27). Rarely, in less than 1% of cases, the right VA may originate from the right common carotid artery or have a common origin with the thyrocervical trunk. The take off of

the left VA is usually situated 35 mm distal to the subclavian origin. In 4% of cases, it comes directly from the aortic arch, between the origins of the left common carotid artery and left subclavian artery. It sometimes shows a common origin with the thyrocervical trunk (2%) or a duplicated origin (1%). The VA may be completely absent (1.5 to 3% (28-30)), in which case it is generally replaced by a persistent congenital carotid-vertebral or carotid-basilar anastomosis. In 90% of cases, it enters the transverse foramen at the C6 level. Less frequently, the level of entry may be situated at C5 (6%) or C7 (3.5%), exceptionally at C4 (1.3%) or C3 (0.1%) (31).

Fenestrations and duplications are relatively rare anomalies that most frequently occur in the vertebral and basilar arteries. Although the terms “fenestration” and “duplication” are sometimes used interchangeably, they represent distinct conditions with fundamentally different mechanisms of formation. Fenestration refers to a division of the arterial lumen, separating the artery into two endothelium-lined channels, which may or may not share their adventitial layer (32) (Fig. 6). Duplications consist in two separate vessels that follow different courses.

The incidence of VA fenestrations varies from 0.19 to 1.9% (Table 3). Most cases have been reported in patients of Japanese origin, which raises the question of ethnic predisposition. Fenestrations generally represent a stable situation and are demonstrated incidentally. The most frequent anomalies associated with fenestrations are aneurysms, but no relation seems to exist between the localization of the fenestrations and the aneurysm (33-37). Other associated anomalies include cerebral arteriovenous malformations (34, 35, 38-40), duplications of the basilar artery (33), occipital-vertebral anastomosis(33), termination of the VA in PICA (33), an accessory middle cerebral artery (37), and malformations of the cervical spine (41) (42). Most, if not all of these associated anomalies are likely fortuitous. The main

differential diagnosis of fenestrations is arterial wall dissection with a double lumen sign. The double lumen results from the parallel patency of the true arterial lumen and of the false lumen located within the arterial wall. This is an important distinction since arterial dissections carry at the acute stage a risk of thrombo-embolic complications that warrants anticoagulation therapy.

Most VA fenestrations are located at the V4 segment and likely form around the hypoglossal nerve. Fenestrations of the cervical VA are extremely rare, and seem to be due to a similar mechanism at the level of cervical spinal nerves.

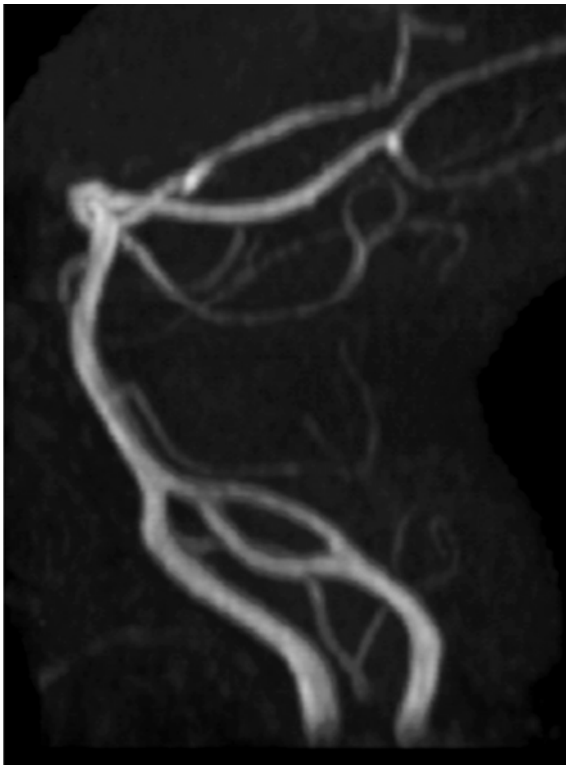


Fig. 6: MRA, showing a fenestration in the left distal VA. Note that both parts of the vessel follow the same course.

Author	Year	No. of cases	Incidence (%)	Identification
Kowada et al. (33)	1972	362	1.90	angiography
Carella et al. (43)	1978	1290	0.23	angiography
Tokuda et al. (44)	1981	300	1.00	angiography
Rieger et al. (42)	1982	500	0.40	angiography
Sanders et al. (32)	1993	5190	0.19	angiography
Mitterwallner (45)	1955	360	0.28	autopsy
Krayenbühl et al. (46)	1957	400	0.25	autopsy
Wollschlaeger et al. (47)	1967	291	0.34	autopsy

Table 3: incidence of VA fenestrations

Duplications of the extracranial VA have traditionally been attributed to variations in the course of the LSA (8). In this variation, one of the vessels forming the duplication (i.e., the LSA) leaves the transverse foramina, enters the spinal canal, and takes a subarachnoid course to rejoin the VA at a different segmental level.

The basilar artery

The basilar artery results from the union of the two VAs at the pontomedullary junction. It courses on the ventral aspect of the brain stem, generally on the midline, and bifurcates into the two posterior cerebral arteries at the level of the pontomesencephalic junction. The basilar artery gives off three types of lateral branches. Paramedian branches are short vessels that penetrate perpendicularly into the pontine parenchyma and supply the medial basal pons. Short circumferential vessels supply the ventrolateral basal pons. The long circumferential arteries include the PICA, the anterior inferior cerebellar artery (AICA), and the superior cerebellar artery (SCA), which are described below.

Basilar artery fenestrations have been reported with a frequency of 0.02-2% (32, 38, 47) in angiographic series. Autopsy series yield higher values (47, 48) (49, 50) (1.3-5.3%), probably because small fenestrations are sometimes not visualized on some angiographic incidences or by less precise imaging techniques. They are generally thought to result from segmental absence of fusion of the two primitive LNA. They frequently coexist with aneurysms of the vertebrobasilar junction (32, 51).

The posterior inferior cerebellar artery

The PICA has the most variable course of the cerebellar arteries. It generally originates from the intracranial portion of the VA, approximately 15 mm proximal to the vertebrobasilar junction, along the lateral aspect of the medulla. From its origin, the PICA courses laterally, along the roots of the hypoglossal nerve, forms a caudal loop over the dorsolateral aspect of the medulla, and turns upward to pass between the tonsil and the vermis, at which point it divides into its medial (vermian) and lateral (hemispheric) branches (Fig. 7).

Lister (52) proposed a division of the PICA into five segments. The first segment, termed *anterior medullary segment*, extends from the origin of the PICA to the inferior olive. The *lateral medullary segment* courses besides the medulla, forming a loop with a caudal or lateral convexity (25), and extends to the origin of the glossopharyngeal, vagal and accessory nerves. The *tonsillomedullary segment* passes posteriorly and then ascends to the medial surface of the tonsilla, forming a loop with caudal convexity (53). The *telovelotonsillar segment* ends at the junction of the vermis, tonsils and hemispheres. The *cortical segment* divides into a medial trunk that supplies the inferior vermis and the adjacent part of the hemisphere, and a lateral trunk destined to the inferior surface of the ipsilateral hemisphere and tonsilla.

The PICA is a late phylogenetic acquisition that appears fully developed only in human (54). Common variants involving the PICA include low origins at C1, C2, or C3, usually explained by variations in size and configuration of the LSA, as well as origin of the PSA from the PICA.

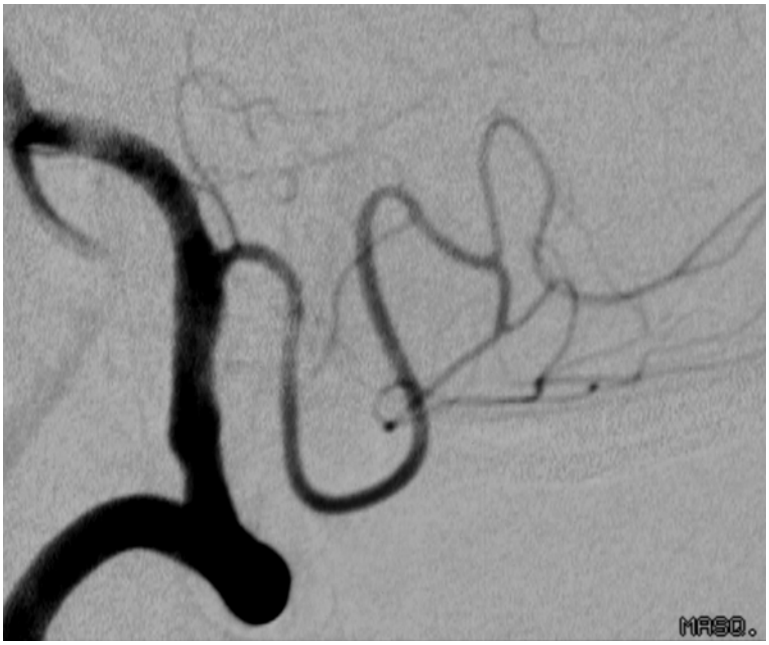


Fig. 7: DSA, left VA injection, lateral view, showing the course of the PICA with its characteristic caudal loop

The anterior inferior cerebellar artery

The AICA generally arises from the inferior third of the basilar artery and courses laterally and slightly inferiorly. At the level of the cerebellopontine angle, it forms a recurrent loop and gives off the internal acoustic artery. This loop not infrequently enters and exits the internal acoustic meatus. It then divides into a lateral trunk that supplies the flocculus and the semilunar lobules, and a medial trunk that feeds the biventral lobule. The AICA may have a

double or triple origin, resulting from separate origins of its medial and/or lateral trunks, and of the internal acoustic artery (55). The AICA may also share a common trunk with one of the other cerebellar arteries.

The superior cerebellar artery

The SCA originates from the rostral end of the basilar artery, near its point of bifurcation. It is traditionally divided into three cisternal segments. The first or *interpeduncular segment* lies on the anterior aspect of the rostral pons and frequently forms a loop that is convex inferiorly. *The ambient segment* courses on the lateral aspect of the pons, paralleling the course of the trochlear or fourth cranial nerve (CN IV), and then turns posteriorly over the middle cerebral peduncle. This segment generally gives off a cortical branch (called lateral marginal branch) and hemispheric branches to the surface of the cerebellum. In its *quadrigeminal segment*, the SCA may lie very close to its contralateral counterpart near the midline. It continues as the superior vermian branch that courses posteriorly over the vermis, close to the midline, and anastomoses with branches of the PICA.

Double or even triple origins of the SCA have been reported, resulting from separate origins of the vermian, hemispheric and lateral marginal branches from the basilar artery. The SCA and more rarely the lateral marginal branch may originate from the posterior cerebral artery (PCA) (55).

The posterior cerebral artery

The PCA arises from the bifurcation of the basilar artery at the pontomesencephalic junction. It can be divided into four segments. The first segment (P1) courses laterally and anteriorly towards the origin of the posterior communicating artery (PComA). The P2 segment begins at the origin of the PComA and turns around the superior cerebral peduncle to enter the ambient cistern. The P3 segment starts at the level of the posterior midbrain, courses in the lateral aspect of the quadrigeminal cistern and extends to the anterior limit of the calcarine fissure. The P4 segment divides into the calcarine and parieto-occipital branches. The PCA gives rise to perforating arteries to the thalamus and circumflex branches to the brain stem.

In about 25% of cases, the PComA is prominent, and the P1 segment is hypoplastic, in which case the PCA is seen arising from the internal carotid artery. This configuration is traditionally but wrongly referred to as the “fetal PCA” (see below).

Persistent carotid-vertebral and carotid-basilar anastomoses

Prior to the establishment of a functional posterior circulation, the vertebrobasilar territory is supplied by the primitive carotid arteries via a series of transverse anastomoses that form and subsequently regress in a caudocranial order (i.e., the trigeminal, hypoglossal, and proatlantal arteries). Persistence of one of these primitive anastomoses into adult life occurs in 0.1% to 1.25% of cases (56-59). The existence of a fourth carotid basilar anastomosis, the otic artery, has been strongly debated. This vessel has never been clearly demonstrated. On the other hand, as it will be shown later, the PComA can truly be considered the cranialmost carotid-basilar anastomosis.

The persistent trigeminal artery

The trigeminal artery is the most frequent persistent carotid-basilar anastomosis, accounting for approximately 85% of cases (59). According to large angiographic studies, its incidence varies from 0.1 to 0.6% (56, 57, 60). The persistent trigeminal artery (PTA) connects the proximal intracavernous portion of the internal carotid artery to the distal basilar artery. It may take a parasellar course, in which case it originates from the posterolateral aspect of the internal carotid artery and courses laterally to the dorsum sellae, and medial to the ophthalmic branch of the trigeminal nerve. Alternatively, it may take an intrasellar course, in its proper canal or groove, in close contact with the pituitary gland. In either case, the artery joins the distal basilar artery between the origins of the AICA and SCA. Blood generally flows in the direction of the basilar artery. In 0.2-0.8% (61-63) of cases, the trigeminal artery ends in one of the cerebellar arteries, and is then referred to as “persistent trigeminal artery variant”. PTAV most frequently end in the AICA or the PICA, less frequently in the SCA(64).

The persistent hypoglossal artery

The hypoglossal artery is the second most common persistent carotid-vertebrobasilar anastomosis, with a reported incidence of 0.03 to 0.26% according to angiographic studies (65, 66). The hypoglossal artery arises from the internal carotid artery, at the level of the superior cervical spine (C1-C3). From its origin it ascends anterior to the VA and the atlas, penetrates the hypoglossal canal medial to the hypoglossal nerve and joins the proximal basilar artery near its origin. According to the diagnostic criteria described by Lie (67) and revised by Brismar (68), 1) the persistent hypoglossal artery leaves the internal carotid artery as a large extracranial branch, 2) It passes through the hypoglossal canal, and 3) The basilar artery originates from the persistent hypoglossal artery.

Associated anomalies

Persistence of carotid-vertebrobasilar anastomoses frequently occurs in association with aplasia or hypoplasia of one or both VAs (46%) or with absence of at least one PComA (8%), reflecting particular constraints during embryogenesis. Numerous other associated anomalies have been reported, although their significance is unclear. They include aneurysms (69) (70, 71) (72-75), cerebral arteriovenous malformations (76-79) (80-82), carotid-cavernous fistulae (83, 84), Moyamoya disease (85, 86) (85, 87, 88), Sturge-Weber syndrome (89) and hemangiomas of the head and neck (90) . Here again, these associations are likely fortuitous.

Clinical implications

Persistent carotid-vertebral anastomoses are generally clinically silent anatomical variations. Isolated cases of oculomotor paresis and trigeminal neuralgia have been reported in association with a PTA (91-93). By analogy, glossopharyngeal neuralgia and hypoglossal nerve palsies have been reported as rare manifestations of a persistent hypoglossal artery (92). However, persistent embryonic anastomoses may be of clinical importance in particular circumstances. In case of carotid stenosis for instance, ischemic lesions in the posterior circulation may occur, either because of reduced blood flow to the vertebrobasilar territory or when embolization through the PTA occurs (94-97). Inversely, collateral flow through a persistent carotid-basilar anastomosis may supplement the carotid circulation in case of a proximal stenosis. Knowledge of the existence of a PTA is useful before surgical interventions in the carotid arterial system or during parasellar surgery, in order to avoid embolization and massive hemorrhage, particularly with the growing popularity of endoscopic neurosurgery. It is equally important when planning endovascular interventions or procedures, such as a Wada test or a carotid artery balloon occlusion test (98).

PART II: DEVELOPMENTAL ANATOMY

THE DEVELOPMENTAL ANATOMY OF THE ARTERIES SUPPLYING THE SPINAL CORD

The embryology of the spinal cord vessels has been studied by Sterzi (99) and by Hoche (100) in animals, and by His (101), Torr (102, 103) and later Zawilinsky (104) in humans. The primitive arterial vascularization of the spinal cord relies on the dorsal aorta, which gives off intersegmental arteries supplying the somites. Each intersegmental artery reaches the anterolateral aspect of the developing spinal cord and establishes longitudinal anastomoses at the capillary level with intersegmental arteries from the levels above and below. At the end of the third week, one can thus distinguish a fine longitudinal capillary network along each anterolateral aspect of the spinal cord. The anterior aspect of the cord situated between the plexuses, and the posterior part of the spinal cord are still avascular at this stage (14). Subsequently, radicular arteries form by the division of the primitive intersegmental arteries into an anterior and a posterior branch. The posterior branches reach the posterior aspect of the cord, and like their anterior homologues, form longitudinal anastomotic networks along the surface of the spinal cord. The posterior radicular arteries give rise to two posterior longitudinal axes on each side. The larger one courses posterior to the dorsal root. The other one, which is finer, forms laterally, between the dorsal and ventral nerve roots, in proximity to the dentate ligament. It is only found in superior vertebrates and humans (14).

With time, the capillary bed over the anterolateral surface of the spinal cord progressively extends to its anteromedian aspect. Between the sixth and tenth weeks of gestation, a single ASA forms. Whether this occurs by fusion of the two parallel anterior axes or by a process of partial obliteration and remodeling of the primitive plexiform anterior axis

is not clear. Incomplete fusion could explain why the anterior spinal axis is often fenestrated and sometimes duplicated, whereas obliteration and remodeling might account for the commonly tortuous course of the ASA (14, 105). By the tenth week, the vasculature of the spinal cord essentially corresponds to its postnatal configuration (104).

After this period, only a few significant changes occur. The posterior anastomotic chains are remodeled into two distinct PSA. There is a significant reduction in the number of radiculomedullary arteries supplying the spinal cord, with a transition from a regular, symmetric and ubiquitous distribution to an irregular and asymmetric distribution (104) (Fig. 8), a transformation that is particularly marked at the level of the inferior third of the spinal cord. The course of radiculomedullary arteries is modified: during embryological development, the unequal growth of the spinal cord and the spinal column results in an apparent ascension of the cord relative to the column, a phenomenon that continues into postnatal life until about the age of twelve months. Consequently, the radiculomedullary arteries assume an ascending course, which becomes more and more marked as one progresses in a caudal direction (106). From the fifth month onwards, the longitudinal spinal axes become tortuous, particularly at the level of the cervical and lumbar enlargements (105-107). They start to straighten around the third year of postnatal life (106).

The number of anterior and posterior spinal contributors detectable by macroscopic anatomy and angiography decreases with age, until the spinal vascularization seems to be derived from a few branches only, including the artery of the lumbar enlargement, or artery of Adamkiewicz. It has been clearly shown, however, that at the mesoscopic and microscopic levels, the anterior and posterior spinal contributors are present at every single level throughout life(108). The apparent vascular distribution observed in the adult human results from the combination of functional adaptation (largest branches at the level of the largest

neuronal masses) and senescence. These observations are clinically important, as they imply that each radicular artery has the potential to become a feeding branch for a vascular malformation.

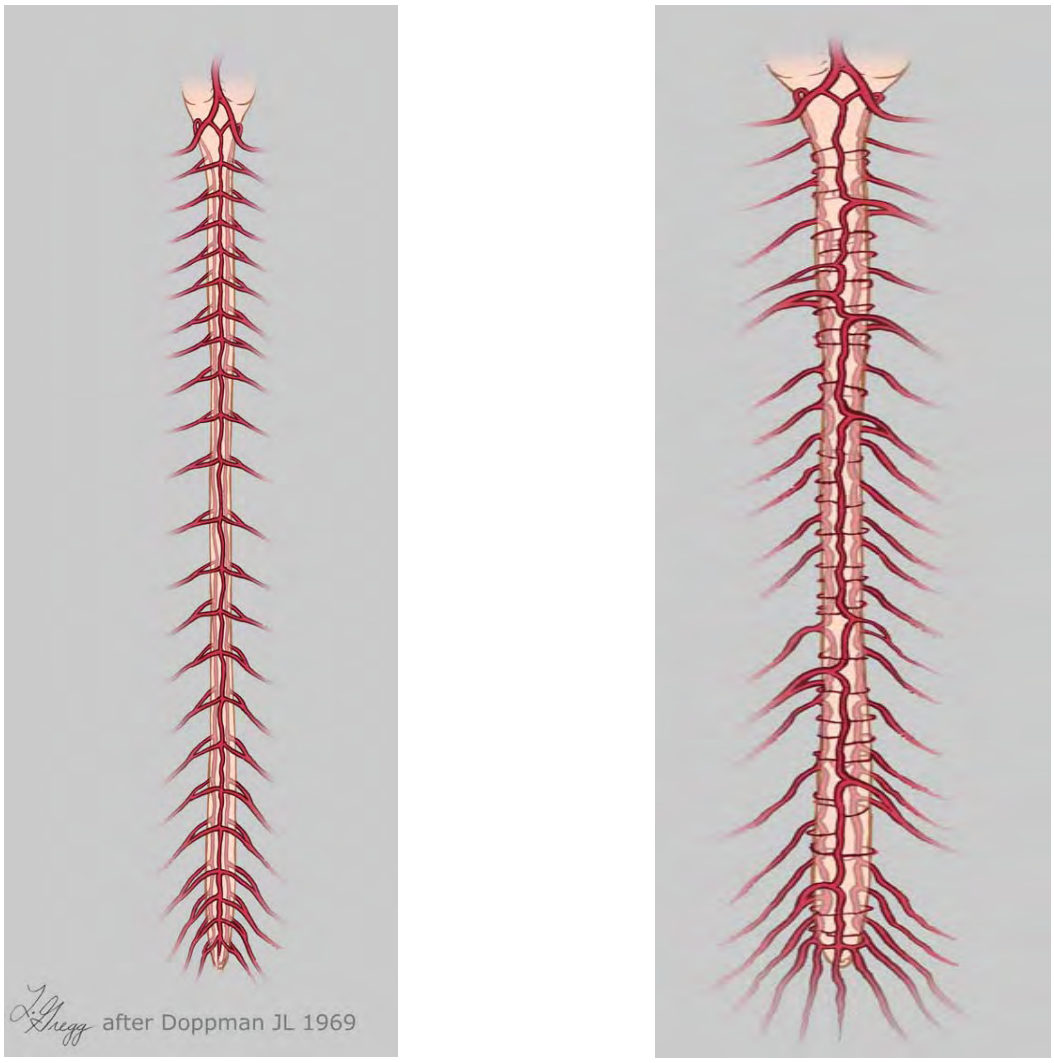


Fig. 8: transformation of the spinal vascularization from a regular pattern, where all radiculomedullary arteries contribute equally to the longitudinal vascular axes (left) to an unequal distribution in the adult configuration (right), resulting from functional adaptation (largest branches at the level of the largest neuronal masses) and senescence

THE DEVELOPMENTAL ANATOMY OF THE VERTEBROBASILAR SYSTEM

The developmental anatomy of the vertebrobasilar system was extensively studied by Padget (4) and by Congdon (5) in humans, and by Moffat (6) in the rat.

The aortic arches

During the fourth week of gestation, the process of embryonic folding carries the two dorsal aortae to a ventral position, a change that results in a dorsoventral loop of their cranial end. The dorsal aortae are initially separate, but subsequently fuse and form a single midline aorta from the fourth thoracic to the fourth lumbar segments. Their unfused cranial ends give rise to a series of bilateral aortic arches that develop within their respective pharyngeal arches. These arches sequentially form and regress, the last ones developing as the first ones disappear. The first aortic arch supplies blood to the primitive carotid arteries. After involution, a portion of it remains as a small branch of the internal carotid artery, the artery of the pterygoid canal or vidian artery. The second arch regresses except for a small remnant that becomes the stapedial artery. The third aortic arch gives rise to the carotid artery. The left fourth aortic arch becomes incorporated in the aortic arch and the proximal descending aorta, whereas on the right side, the fourth aortic arch contributes to the formation of the proximal subclavian artery. The sixth arch is at the origin of the pulmonary arteries and *ductus arteriosus*.

The primitive carotid arteries

In parallel with the brain development, the cerebral arterial tree first proliferates into an anterior circulation first, and later into a posterior circulation as well. The primitive internal carotid arteries develop mainly from the third aortic arches and have two principal divisions. The cranial division supplies the developing forebrain and gives rise to the anterior cerebral, middle cerebral, and anterior choroidal arteries. The caudal branch corresponds to the PComA, which supplies the posterior choroidal, diencephalic and mesencephalic arteries. However, the PComA should in fact be considered as the last carotid-basilar anastomosis (see below).

The primitive vertebrobasilar system

The posterior circulation initially consists of two parallel longitudinal vascular channels (i.e., the LNA), which later fuse over the midline to form the basilar artery. Prior to the development of the VA, the posterior circulation is supplied by a series of transverse pre-segmental anastomoses originating from the carotid circulation. These anastomoses include the posterior communicating artery (also labeled as the caudal division of the internal carotid artery in early embryonic stages), the trigeminal artery, the hypoglossal artery, and the pro-atlantal artery of Padget (ProA) [1]. They follow a complex pattern of formation and regression. As it will be discussed in more details later, our findings, supplemented by the work of previous authors(7) (109) strongly suggest that the PComA should be considered the last carotid-basilar anastomosis rather than the caudal division of the primitive carotid, as generally believed.

[1] The existence of a significant primitive otic artery in man remains debated.

The ProA is located between the most caudal pre-segmental artery (i.e., the hypoglossal artery) and the first intersegmental artery (i.e., the artery accompanying the second cervical nerve); it courses between the occipital bone and the atlas, along with the first cervical nerve. Its branching pattern closely resembles the typical distribution of spinal radicular arteries. In particular, it gives off a dorsospinal division that further divides into a ventral and a dorsal branch. The ventral branch supplies the developing basilar circulation and eventually becomes incorporated into the definitive VA. In addition, the ventral branch gives off an ascending and a descending branch that later fuse with their contralateral counterpart to give the ASA. Unlike the spinal branches at other cervical levels, the spinal branch of the ProA and its ventral radicular component remain prominent in the adult; in fact, they eventually form the distal segment of the definitive VA. The dorsal branch courses along the posterior nerve root. Like its ventral homologue, it divides into ascending and descending branches. The ascending branch establishes a connection with the PICA, whereas the descending branch anastomoses with the posterior spinal plexus (the future PSA) (6), and later becomes the trunk of origin of the PSA. The left and right PSAs do not fuse, but remain separate vessels on each posterior-lateral aspect of the spinal cord.

The definitive VA takes form through a series of longitudinal anastomoses linking the first six cervical intersegmental arteries (5). Because of its peculiar mode of formation, the VA has been described as a “hemodynamic solution” rather than a true vessel (55). Eventually, the proximal connections of the intersegmental arteries to the dorsal aorta regress, at the exception of the sixth intersegmental artery, which remains prominent and becomes the adult subclavian artery (110). The longitudinal connection between the first intersegmental artery and the ProA establishes the continuity between the subclavian artery and the basilar artery. Although the proximal trunk of the ProA involutes, its dorsal branch becomes a portion of the adult VA (*i.e.*, the distal VA and its so-called C1 muscular branch). Through

this assimilation of the distal ProA by the VA, the cranial origins of the ASA and PSAs, which originally correspond to the anterior and posterior radicular branches of C1, become distal branches of the adult VA.

The posterior communicating and the posterior cerebral arteries

The PComA develops as a caudal branch of the internal carotid artery and supplies the LNA as well as the posterior choroidal and diencephalic arteries. As mentioned above, it should be considered as the last of the carotid-basilar anastomoses providing flow to the developing posterior circulation, but unlike the other anastomoses, the PComA generally persists into adult life.

The territory of the PCA is initially supplied by the anterior choroidal artery, which is, as stated above, a cranial branch of the internal carotid artery (Fig. 9). The anterior choroidal artery eventually anastomoses with the posterior choroidal artery (a branch of the PComA) and thus establishes the continuity between the developing basilar artery and the future PCA. Later, the anterior choroidal artery looses its connection to the developing PCA, so that the PCA territory is completely taken over by the vertebrobasilar system. The origin of the PCA divides the PComA into a proximal and a distal segment, the latter being traditionally referred to as the “P1 segment” of the adult PCA. From an embryonic standpoint, however, this clearly represents a misnomer, as the so-called “P1 segment of the PCA” is not derived from the fetal PCA, but from the PComA.

Different arterial configurations may result, depending on the relative development and regression of the two segments of the PComA. The distal segment generally becomes prominent compared to the proximal one, so that the PCA is seen arising as a branch of the

basilar artery. When, however, the proximal segment remains prominent, the PCA appears as a continuation of the PComA, a configuration that is classically referred to as the “fetal PCA”, another misnomer. This term should be reserved to variants characterized by a prominent anterior choroidal artery that supplies the PCA territory. In the second part of this work, we will provide examples of these different arterial configurations, and discuss their mechanism of formation

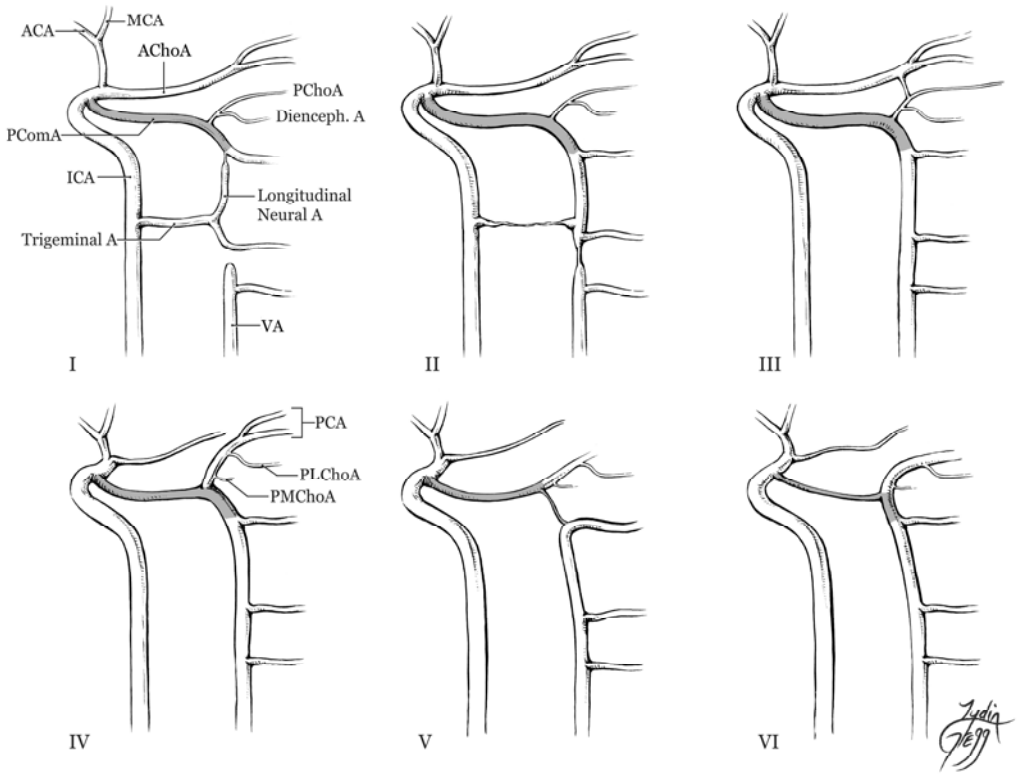


Fig. 9: developmental anatomy of the distal basilar artery and posterior communicating artery complex

I, The anterior circulation supplies the developing posterior circulation, i.e., the parallel longitudinal neural arteries (LNA), via the trigeminal and the posterior communicating arteries. The terminal branches of the carotid axis are the anterior choroidal artery (AChoA), which provides the territory of the posterior cerebral artery, and the anterior cerebral artery (ACA) and its developing branch, the middle cerebral artery (MCA). The VA is not yet connected to the LNA. The PComA has two small branches, the posterior choroidal artery (PChoA) and the diencephalic artery.

II, The VA connects to the proximal end of the LNA/basilar artery. The trigeminal artery involutes, the PComA becomes the last of the carotid basilar anastomoses.

III, A connection is formed between the AChoA/PCA and the posterior choroidal artery (PChoA).

IV, The connection between the PComA and the PCA is now well established. The AChoA is not anymore supplying the PCA territory. The latter can be supplied equally by the posterior circulation via the distal segment of the PComA, or by the anterior circulation via the proximal segment of the PComA. This configuration of equal contribution can persist at the adult stage. Other adult configurations depend on the relative regression of the PComA, as described below (V and VI). Note that the diencephalic branch is now called the posterior medial choroidal artery (PMChoA), while the original posterior choroidal artery (PChoA) is now called the posterior lateral choroidal artery (PLChoA). Although both arteries are originally derived from the PComA, they are considered as branches of the PCA at the adult stage. The so-called “P1 segment of the PCA” is in fact the distal portion of the PComA.

V, In this second adult configuration, the distal segment of the PComA has regressed and the anterior contribution to the PCA becomes dominant. This configuration is often mislabeled as a “fetal origin of the PCA”. The true “fetal origin of the PCA”, where the transfer of the PCA territory from the AChoA to the PComA has not occurred, is not illustrated here, but would be similar to scheme II. This latter configuration is for example the normal adult anatomy in birds.

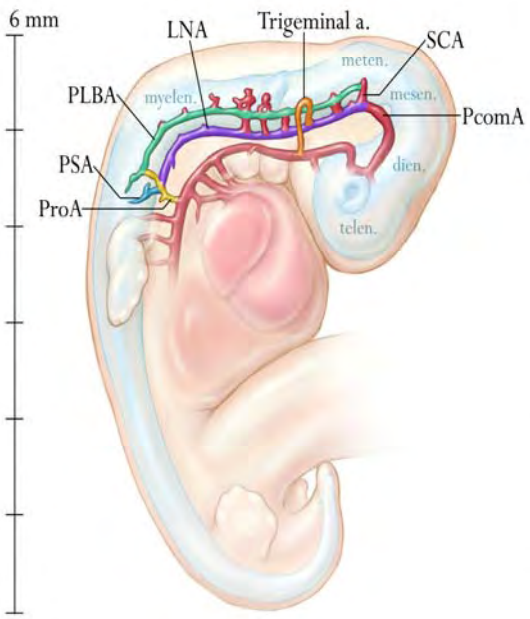
VI, In this third adult configuration, the proximal segment of the PComA has regressed and the posterior contribution to the PCA becomes dominant.

The primitive lateral basilovertebral anastomosis of Padget

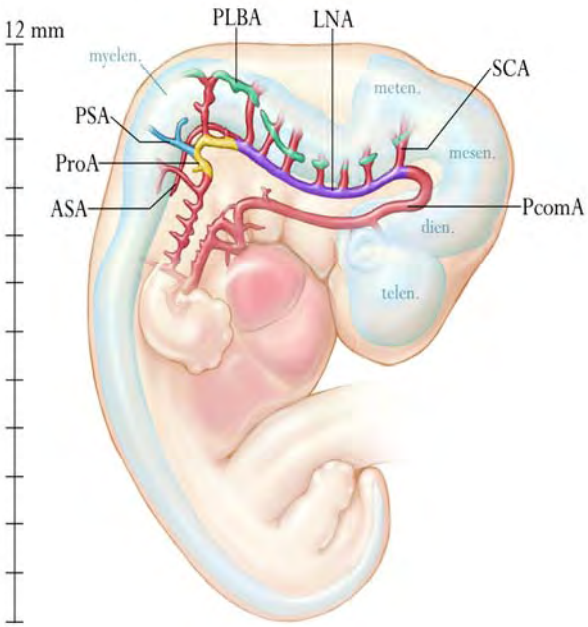
The primitive lateral basilovertebral anastomosis of Padget (PLBA) (4) is an embryonic structure that originates as a cranial branch of the ProA (6, 110) and parallels the developing basilar artery (i.e., the longitudinal neural plexi or LNA), coursing between the sixth and twelfth cranial nerves medially, and the seventh to eleventh nerves laterally (Fig. 10). The PLBA anastomoses with lateral branches of the LNA, some of which will later become the proximal segments of the cerebellar arteries. Although pre-segmental arteries such as the trigeminal artery are most commonly connected to the LNA, Moffat has shown in the rat the existence of direct anastomoses between pre-segmental branches and the PLBA, without connection to the basilar artery (6). The PLBA lies on the same longitudinal axis as the LSA. The PLBA and the LSA thus respectively constitute the intracranial and extracranial portions of the same vascular channel. This correspondence was clarified by Moffat (6) in his studies on the development of hindbrain arteries in the rat, a species in which the PLBA persists into adult life and the continuity of the lateral axis is readily apparent [2]. Padget (4) anticipated that variations of the VA and its branches resulted from partial persistence of the PLBA. In the second part of this work we will confirm this hypothesis, and illustrate it with several vertebrobasilar variations.

[2] Although the PLBA is named “lateral longitudinal artery” in his work, Moffat confirmed the equivalence of the two structures.

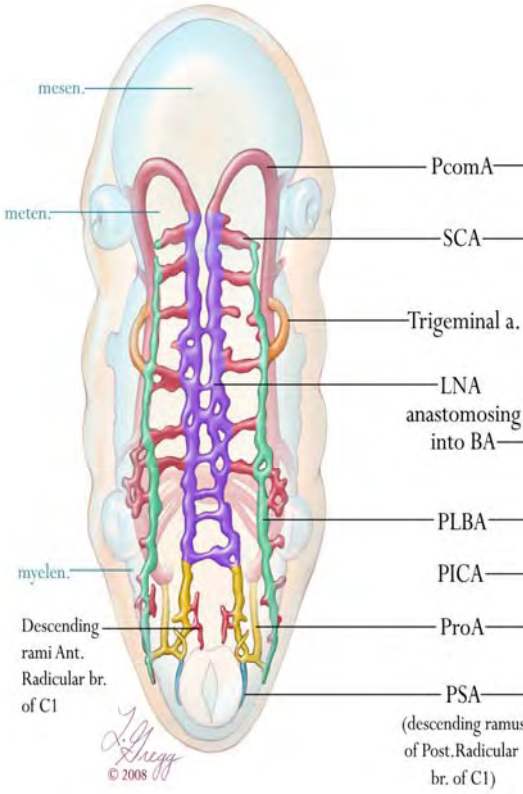
Carnegie Stage 14, (5-7mm) lateral



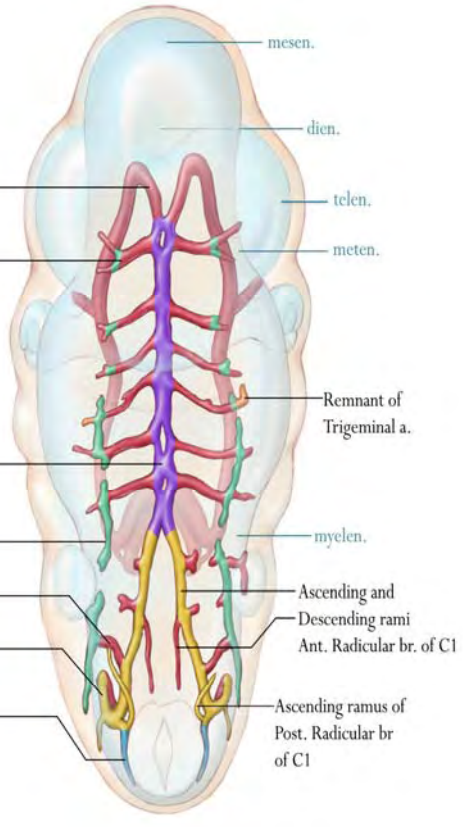
Carnegie Stage 17, (11-14mm) lateral



Carnegie Stage 14, (5-7mm), superior-dorsal



Carnegie Stage 17, (11-14mm) superior-dorsal



PcomA
SCA
Trigeminal a.
LNA anastomosing into BA
PLBA
PICA
ProA
PSA (descending ramus of Post.Radicular br. of C1)

Remnant of Trigeminal a.
Ascending and Descending rami Ant. Radicular br. of C1
Ascending ramus of Post. Radicular br of C1

Fig. 10. The primitive lateral basilovertebral anastomosis (PLBA) of Padget

Representation of the cranial arterial system at the developmental stages 2 (left) and 4 (right), seen from lateral (top) and dorsal views (bottom). The vessels are seen through the body of the embryo. The PLBA is highlighted in green. The PLBA is considered to be a branch of the ProA (yellow) coursing cranially lateral to the LNA (purple), to which it is connected by several transverse anastomoses. Some of these anastomoses will later become the proximal segments of the cerebellar arteries as well as other branches of the basilar artery. The descending (or extracranial) portion of the PLBA corresponds to the adult lateral spinal artery (LSA). The ProA (of Padget) is the radicular artery accompanying the first cervical nerve. Its anterior radicular branch divides into ascending and descending rami that will respectively become the vertebrobasilar junction and the anterior spinal artery after fusion with their contralateral homologue. The posterior radicular branch of the ProA also divides into ascending and descending rami, but these do not fuse with their contralateral homologues. The descending ramus becomes the posterior spinal artery, while the ascending ramus establishes a small anastomosis with the ipsilateral PICA. This anastomosis can sometimes become prominent, explaining the apparent origin of the PICA from the extracranial vertebral artery at C1 (“PSA-type”). The connection between the PLBA and its extracranial equivalent (LSA) explains, on the other hand, the second type of C1 origin of the PICA (“LSA-type”).

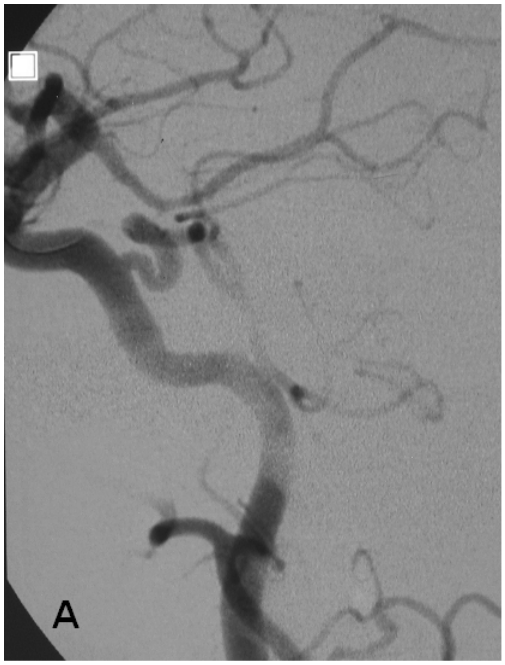
PART III: VERTEBROBASILAR VARIATIONS AND THEIR MECHANISMS OF FORMATION

BILATERAL SEGMENTAL AGENESIS OF THE VERTEBROBASILAR JUNCTION

Absence or hypoplasia of the terminal portion of one VA is a commonly observed anatomic variant. In such instance, the VA either shows a very small connection to an otherwise normal basilar artery, or continues its course as the PICA. In both cases, the basilar artery is normal and receives most or all of its blood supply from the contralateral VA. Absence of the terminal portion of both VA is, on the other hand, exceptional. In the following section, two angiographic observations of bilateral absence of the distal VA are illustrated. Remarkably, in both cases, the proximal portion of the basilar artery is missing as well, while the remaining segment of the basilar artery is connected to the anterior circulation through a persistent embryonic vessel. These cases illustrate a congenital anatomic variation that can be explained by the developmental history of the vertebral and basilar arteries. A case of vertebrobasilar junction disease that presents similarities with this anatomic variation is also reported, and their distinguishing features are discussed.

Illustration of cases

Case 1 - A 69-year-old woman was investigated for a right pulsatile tinnitus. Her history was remarkable for arterial hypertension, migraine headaches, and premature birth (30-week gestation). MR imaging and angiography revealed a left PTA possibly associated with an aneurysm. Digital subtraction angiography (DSA) showed no aneurysm, but documented several anatomic variations. Both PCA originated from the distal internal carotid artery (ICA), without detectable connection with the basilar artery. There was no evidence of connection between the VA and the basilar artery on either side. The right VA terminated as a branch of the occipital artery, while the left VA crossed the midline to continue as the right PICA. The left PICA was provided by the left AICA through a common stem (AICA-PICA trunk). Because of the absence of connection with both PCA and both VA, the basilar artery was isolated from its usual sources of blood supply, and was vascularized by the left PTA only (Fig. 10A and B). Finally, the ASA originated from the right inferior thyroid artery (Fig. 10C).



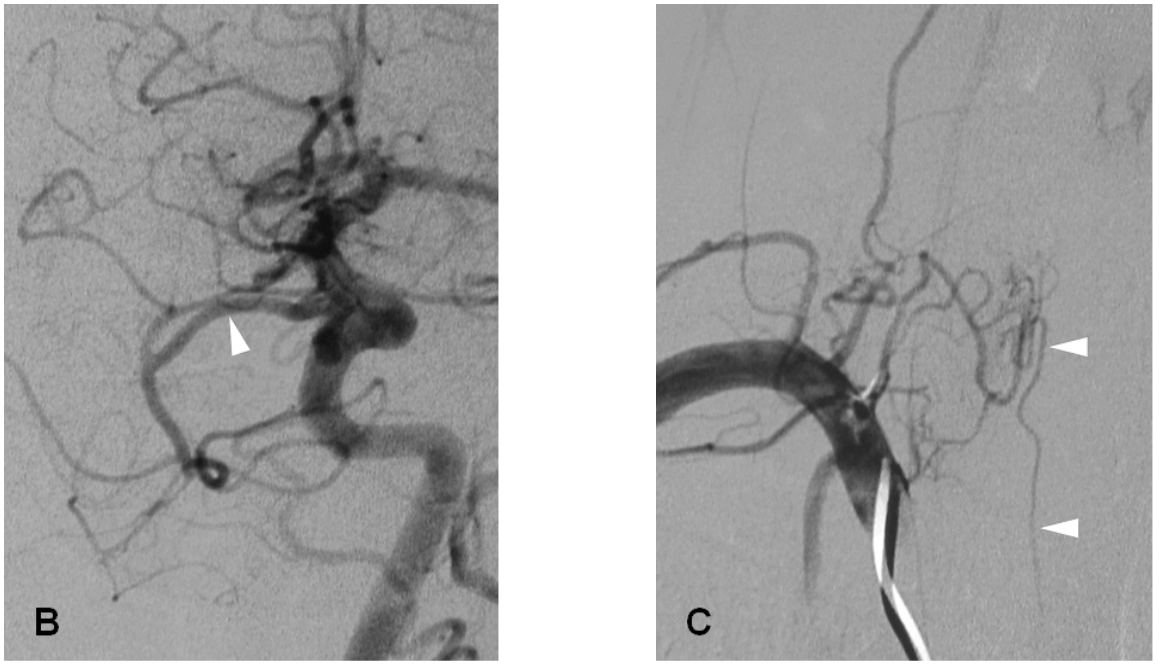


Fig. 10 (Case 1): 69-year-old woman with pulsatile tinnitus.

A, DSA, left common carotid injection, right anterior oblique view, showing the basilar artery fed by the PTA, and the bilateral absence of connection between the basilar and posterior cerebral arteries. The basilar artery ends into the superior cerebellar arteries distally, and into prominent anterior-inferior cerebellar (AICA) arteries proximally. The left AICA also supplies the left PICA territory. The right PICA territory is fed by the left VA (not shown). There is no visible connection between the VAs and the basilar artery.

B, DSA, left common carotid injection, right anterior oblique view, showing the basilar artery fed by the PTA (arrowhead), and the absence of P1 segment bilaterally. The left AICA also supplies the left PICA territory, while the right PICA territory is fed by the left VA (not shown). There is no connection detected between the VAs and the basilar artery.

C, DSA, right subclavian injection, showing opacification of the ASA (arrowheads) via the right inferior thyroid artery.

Case 2 - A 65-year-old man followed for Wegener granulomatosis and right ear deafness underwent MR imaging and angiography that revealed a possible right ICA termination aneurysm. DSA showed no aneurysm, but documented several vascular variations. On both sides, the VA continued as a PICA without connection to the basilar artery. A prominent right anterior choroidal artery was supplying the territory of the right PCA, without connection to the basilar artery (Fig. 11A). The right PComA, separate from the PCA, was supplying the basilar artery, which showed retrograde opacification down to the level of origin of both AICA (Fig. 11B). Again, there was no connection between the right PComA and the right PCA itself. The left PCA originated from the distal left ICA without detectable connection to the basilar artery. Because of the absence of connection with both PCA and both VA, the basilar artery was isolated from its usual sources of blood supply, and was fed through the right PComA only.

Case 3 - A 51-year old man with a history of coronary artery disease, arterial hypertension, hyperlipidemia, and smoking presented with a one-year history of episodic dizziness associated with a throbbing sensation in both ears elicited by head rotation to the right or left. Initial investigation for benign paroxysmal positional vertigo remained inconclusive. MR imaging and angiography only offered poor visualization of the basilar artery and VA. DSA showed that the extracranial portion of both VA was of small caliber but otherwise unremarkable. The right VA was not connected to the basilar artery, but continued as the vermian branch of the right PICA, while the hemispheric branch took origin from the right AICA. The V4 segment of the left VA, although small and irregular, was seen joining the proximal basilar artery, which showed antegrade opacification up to the level of origin of the left AICA. The vermian and hemispheric branches of the left PICA were both arising from the left V4 segment. The left ICA contributed to the basilar artery via a large PComA connected to the left PCA. See Fig. 12 A-C.

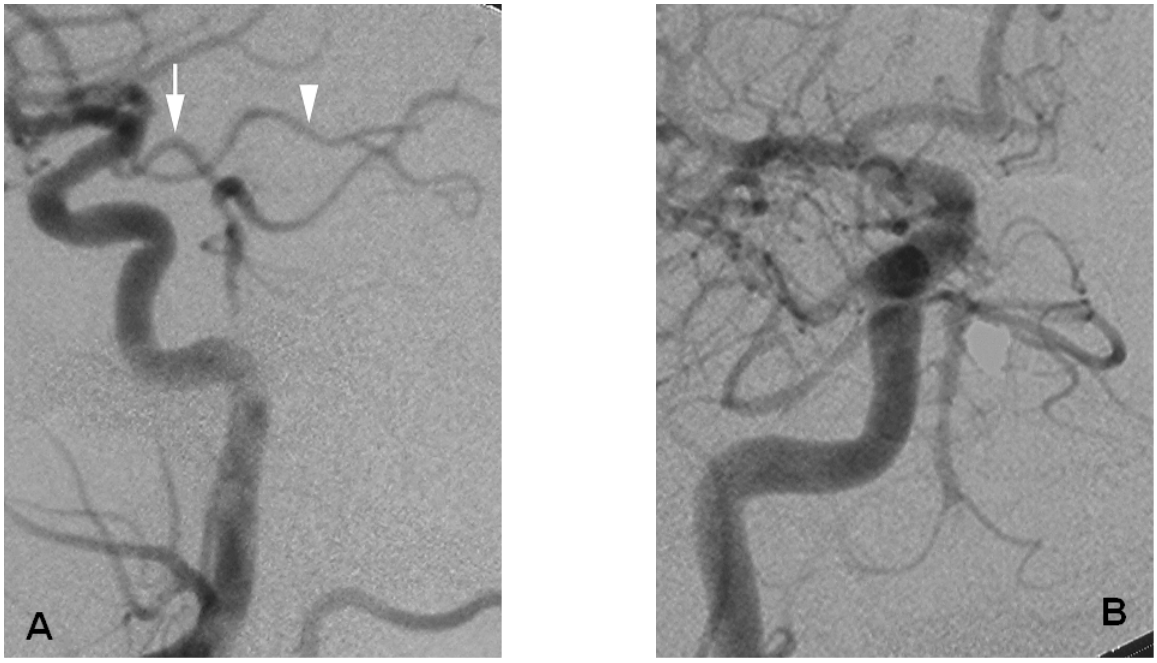
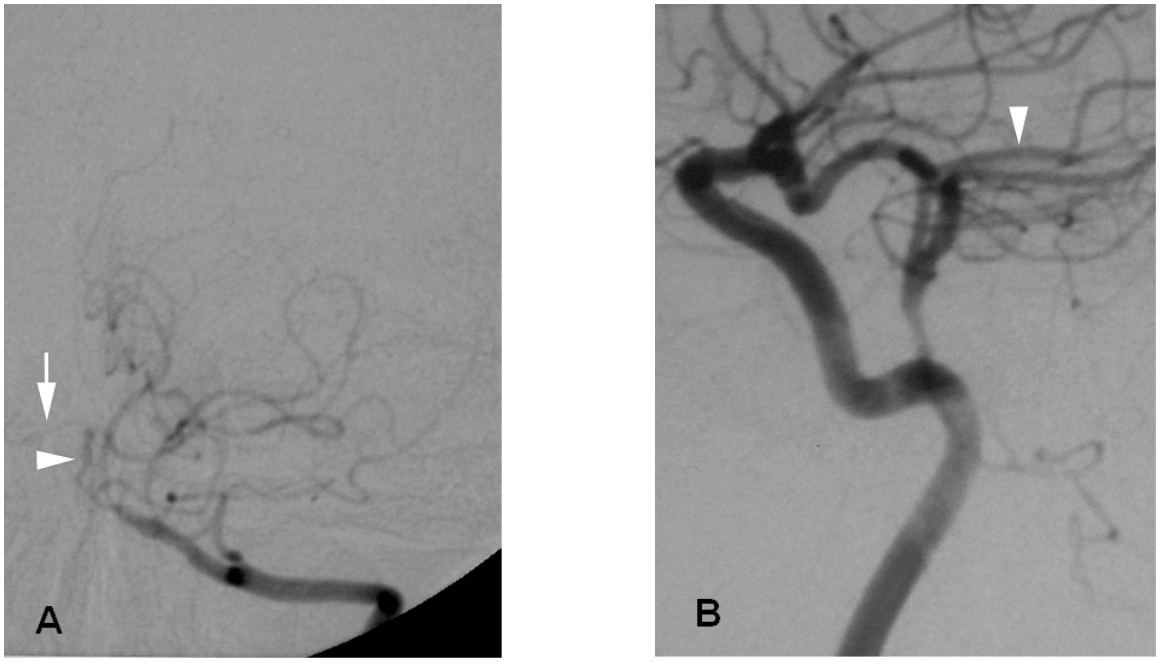


Fig. 11 (Case 2): 65-year-old man with Wegener granulomatosis

A, DSA, right common carotid injection, lateral view, showing a prominent anterior choroidal artery supplying the right PCA territory (arrowhead). Note a separate right PComA feeding the basilar artery (arrow).

B, DSA, right common carotid injection, transfacial view, showing the basilar artery fed by the right PComA. The basilar artery ends into the superior cerebellar arteries distally, and into the anterior-inferior cerebellar (AICA) arteries proximally. Both VAs end as ipsilateral posterior-inferior cerebellar arteries without connection with the basilar artery (not shown).



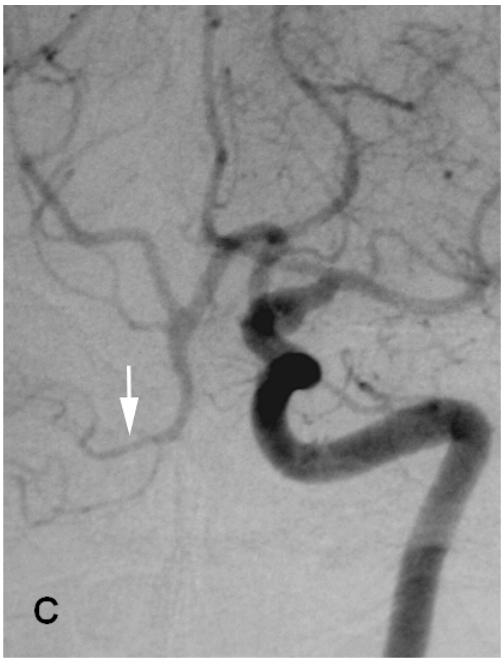


Fig. 12 (Case 3): 51-year-old man with intermittent dizziness

A, DSA, left VA injection, AP view, showing disease in the distal VA and proximal basilar artery (arrowhead), including faint opacification of the right AICA (arrow). **B**, DSA, left common carotid artery injection, lateral view, showing a large PComA supplying the basilar artery. Note that the PComA then continues as the left PCA (arrowhead). The hemispheric branch of the right PICA territory is fed through a connection with the basilar artery. **C**, DSA, left common carotid injection, transfacial view, showing a large left PComA supplying the basilar artery, and continuing its course as the left PCA. Note the opacification of the right AICA (arrow), which confirms the existence of a connection between the left VA and the distal basilar artery.

Discussion

Bilateral vertebrobasilar aplasia

The two anatomic variants presented here are characterized by a set of angiographic findings that include the bilateral absence of connections between the basilar artery and the PCA cranially, and between the basilar artery and the VA caudally. As a result, the basilar artery is, in both cases, isolated from its usual sources of blood supply, in a manner reminiscent of its embryonic state before the connections with the PCA and the terminal VA are established. It is therefore not surprising to find that the blood supply to the basilar artery is provided, in these patients, by a persistent embryonic vessel, *i.e.*, a PTA in Case 1 and a PComA in Case 2. The role played by persistent carotid-basilar anastomoses, in particular by PTA, as sources of collateral supply in cases of congenital anomalies of the carotid and vertebral axes has been well described (111, 112). As mentioned before, the PComA should be considered, in the fetus, as the most cranial of the carotid-basilar anastomoses rather than as the caudal division of the fetal ICA (113). In a superb study dealing with the development of the PCA in the rat, Moffat has shown that the territory of the PCA is first provided by the anterior choroidal artery (not by the PComA, as often believed), and later transferred to the cephalic end of the ipsilateral LNA (*i.e.*, the future) through the formation of a connection with the PCA (7). At the adult stage, the PCA territory is either taken in charge by the BA or by the PComA, depending on the relative degree of regression of the PComA proximal and distal to the connection established with the PCA. This developmental pattern is consistent with modern observations made on human fetuses (109). It therefore appears that the segment of PCA located, in the adult configuration, between the PComA and the basilar artery, and classically named the P1 segment of the PCA, is in fact a portion of the PComA located distal to the newly established connection with the PCA. It also becomes clear that, at the adult stage, a dominant PComA taking in charge the PCA territory should not be called a fetal origin of the

PCA. This appellation should be reserved for the less common variant that sees the PCA territory provided by the anterior choroidal artery. The second case offers a “snapshot” of this early developmental stage, in which the right anterior choroidal artery continues as the PCA, while a separate right PComA connects to the BA as a typical carotid-basilar anastomosis. In this case, the two vessels can be clearly labeled since they are simultaneously present. The distal point of origin of the other three PCA seems compatible with this variant as well, *i.e.*, an anterior choroidal artery origin of the PCA, but definitive identification is in these cases hindered by the lack of a detectable PComA. The absence of connection between the basilar artery and the PCA in each instance is consistent with the hypothesis that the PCA territory has not been transferred from the anterior choroidal artery to the LNA. Again, it is important to realize that the presence or absence of a so-called P1 segment located between the PComA and the basilar artery is irrelevant, since this segment in fact corresponds to the distal portion of the PComA. The vascular segment important to this discussion, whose presence or absence decides of the fate of the PCA territory, becomes at the adult stage a portion of PCA located distal to its connection with the PComA.

Another important observation made in both cases is the absence of the proximal portion of the basilar artery in addition to the bilateral lack of distal VA. This finding comes in fact as a logical consequence of the developmental anatomy of the vertebrobasilar junction. As mentioned above, the proximal basilar artery results from the fusion of the ascending rami of the anterior radicular branches of C1, originating from the left and right ProA. Bilateral aplasia of these anterior radicular branches therefore explains the absence of the proximal basilar artery as well as the absence of both distal VA (Fig. 13). On the other hand, aplasia of a single anterior radicular branch would only result in the absence of one distal VA, the contralateral VA and the basilar artery being unremarkable, a variant that is in fact commonly observed.

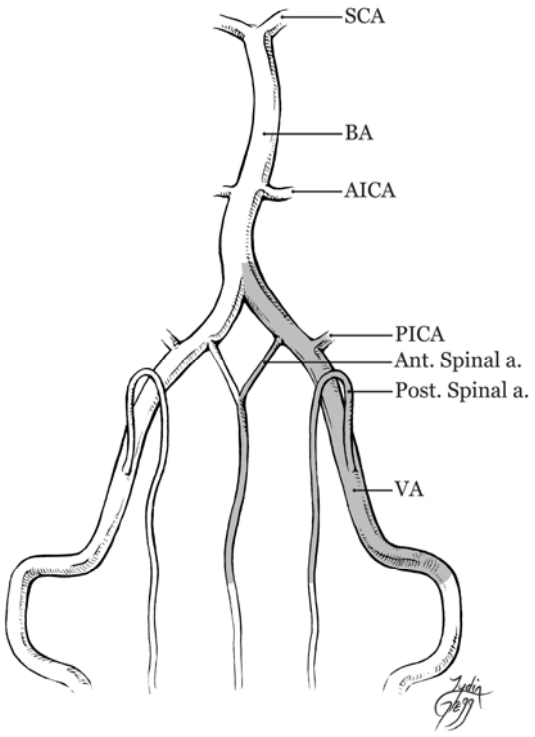


Fig. 13: Schematic representation of the vertebrobasilar circulation, anteroposterior view of the vertebral and basilar arteries. On the left side, the segments derived from the radicular branch of the ProA are shown in grey.

The PICA normally arises from the portion of the VA that is missing in the two patients. The origin of the PICA in these cases documents the utilization of previously described collateral pathways. In the first patient, the left PICA comes from the basilar artery as a common stem with the left AICA (AICA-PICA trunk), while the right PICA is the continuation of the left VA. In the second patient, each PICA is the cranial continuation of the ipsilateral VA. A proximal origin of the PICA from VA can be understood either as a variation of the LSA (8), or as a variation of the PSA (47, 114). In the latter instance, the PICA is a branch of the PSA via the connection normally established between these two vessels by the ascending ramus of the posterior radicular branch of C1. This suggests that the

cause of bilateral vertebrobasilar junction aplasia may be limited to the congenital absence of the anterior radicular branches of C1.

Mechanism of formation

It is conceivable, although not proven, that the absence of connection between the basilar artery and the PCA comes, in our two cases, as a secondary consequence of the lack of proximal connection with the VA and the resulting paucity of antegrade flow within the basilar artery. Alternatively, the concurrent lack of connections between the basilar artery and the PCA cranially, and the basilar artery and VA caudally, could be related to a common, yet unknown mechanism interfering with the development of such vascular segments at a specific moment of the fetal life. A mechanism involving the secondary regression of previously existing vascular segments seems unlikely in view of the well defined, “segmental” pattern of the vascular anomaly, and the persistence of an embryonic branch as source of collateral blood supply. These characteristics may be compared to the case of bilateral regression of the carotid and vertebral arteries reported by Mahadevan and co-authors, in which collateral supply was provided by the development of a vascular rete rather than by an embryonic artery (115).

Finally, in the first patient, the anterior spinal axis receives a prominent contribution from the right inferior thyroid artery. This variant is rare (116). It may be seen here as a collateral pathway developed in order to palliate for the absence of cranial contributors of the anterior spinal axis from the distal VA, *i.e.*, the descending ramus of the anterior radicular artery of C1.

A review of the literature revealed only one observation similar to the variant presented here. Hoh and co-authors have recently described a case of “persistent axial nonfusion of the basilar artery”. In this case, as in the first patient, each VA continues as the ipsilateral PICA and the basilar artery, which is supplied by a PTA, seems absent below the level of the AICA (117). The authors attribute the vascular anomaly to “a failure of fusion of the distal basilar artery”, a “mid-basilar agenesis”, and to “the PICA termination of the VA”. Their variant is similar to the ones documented above. It can be explained by the developmental mechanism described above, *i.e.*, the aplasia of the radicular artery of C1, or of its anterior radicular branch.

Clinical implications

The two patients with vertebrobasilar aplasia described herein were investigated for an unrelated reason, *i.e.*, a pulsatile tinnitus in the first patient and Wegener granulomatosis in the second. In each case, MR angiography raised a suspicion of intracranial aneurysm, which led to the obtainment of a diagnostic angiogram. There was no sign or symptom that could be directly referred to the vascular anomaly in either patient. It can therefore be said of their anatomic variants that they were discovered incidentally. This is not the case of the third patient, whose clinical presentation was suggestive of vertebrobasilar insufficiency. The morphology of the vascular anomalies revealed by DSA in this case was, moreover, pointing towards a pathologic process rather than a congenital variant. The principal feature that helps distinguish the two situations is the presence of a connection between the left VA and the basilar artery. The small and irregular aspect of the distal left VA was consistent with an atheromatous lesion, while the proximal portion of the basilar artery, although abnormal in appearance, was present. The fact that the left PComA, in addition to providing collateral supply to the basilar artery, was in charge of the ipsilateral PCA territory is of particular

interest in regard to the pattern of development discussed earlier. It indicates that the fetal transfer of the PCA territory from the Anterior choroidal artery to the PComA did occur normally in this patient, as opposed to the previous two cases. Identifying the congenital variant (presented in the first two cases) as such might limit unnecessary diagnostic procedures, while recognizing the acquired nature of bilateral aplasia of the vertebral artery (as the presented in the third case) has direct therapeutic consequences.

Conclusion

In summary, two cases of selective bilateral aplasia of the vertebrobasilar junction are presented. This anatomic variation is characterized by the absence of the distal portion of both VA as well as the absence of the proximal portion of the basilar artery. The lack of cranial connection of the basilar artery with the PCA further isolates this vessel from its normal sources of blood supply. As a consequence, the basilar artery is taken in charge by a persistent carotid-basilar anastomosis such as a PTA or a PComA. The mechanism underlying this variant may be as limited as the aplasia of the anterior radicular artery of C1, a branch of the ProA that normally becomes the distal segment of the VA and the proximal basilar artery at the adult stage. The lack of cranial connection of the basilar artery with the PCA could be understood as a remote consequence of the proximal vertebrobasilar aplasia and the resulting paucity of flow within the basilar artery. Alternatively, it is possible that a similar, yet unknown mechanism is responsible for the absence of both the proximal and distal connections of the basilar artery. From a clinical standpoint, this vascular anomaly was discovered incidentally in the two patients, a finding consistent with the supposed congenital nature of the anatomic variant.

DEVELOPMENTAL ANATOMY OF THE DISTAL VERTEBRAL ARTERY IN RELATION TO VARIANTS OF THE POSTERIOR AND LATERAL SPINAL ARTERIAL SYSTEMS

Although the LSA was already identified by Kadyi in 1889 (3), its role in the developmental anatomy of the distal VA was only recognized later by Lasjaunias and colleagues (8). These authors showed, in particular, that anatomic variants such as the C1 and C2 origins of the PICA, the duplication of the VA, or the intradural course of the distal VA, are related to variations in size and connection of the LSA.

In this section a series of anatomic variants of the distal VA and PICA origin are presented. They are similar but not identical to the variants linked to the LSA system, and result from variations in the development of the PSA rather than the LSA. In order to illustrate the analogy between the two types of variants, and highlight some of their distinguishing features, each illustrated variant linked to the PSA system is matched with an equivalent variant derived from the LSA system.

Illustration of cases

The case histories are arranged according to the type of anatomic variation described with, for each type, examples involving the PSA and LSA systems.

C1 origin of PICA

Case 4 – A Wada test was performed in a 20-year-old woman as part of the pre-surgical workup of a left frontal lobe neoplasm. Cerebral DSA incidentally documented a C1 origin of the right PICA. This variant is linked to the PSA (Fig. 14 A and B).

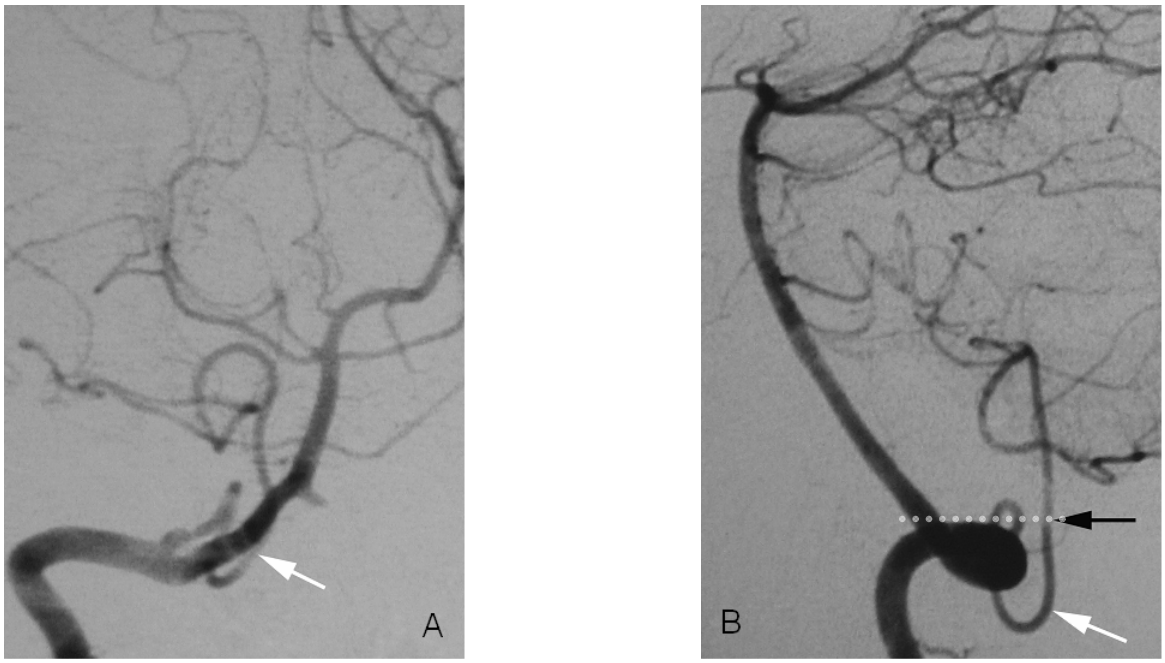


Fig. 14 (Case 4): A proximal origin (C1) of the right PICA (PSA type) in a 20-year-old woman.

A, DSA, right VA, anteroposterior view, showing a proximal origin of the right PICA from the superior aspect of the right VA. The PICA trunk has a Z-shaped course consistent with the normal course of a PSA. From a developmental perspective, the segment of the VA proximal to the take-off of the PICA corresponds to the spinal branch of the ProA, which divides into an anterior radicular branch (the distal VA), and a posterior radicular branch (the PSA). In this case, the ascending ramus of the PSA (white arrow), which normally only establishes a small anastomosis with the PICA, is prominent and constitutes the actual origin of this vessel.

B, DSA, right VA, lateral view, showing the dorsal position of the proximal PICA (black arrow), in the foramen magnum region (whose approximate anterior-posterior diameter is indicated by a dotted line) aligned with the ascending ramus of the PSA (white arrow).

Case 5 – A 15-year-old boy underwent preoperative embolization of a nasopharyngeal angiofibroma. Incidentally, a proximal origin of the right PICA from both C1 and C2 roots was documented. This variant is linked to the LSA (Fig. 15 A and B).

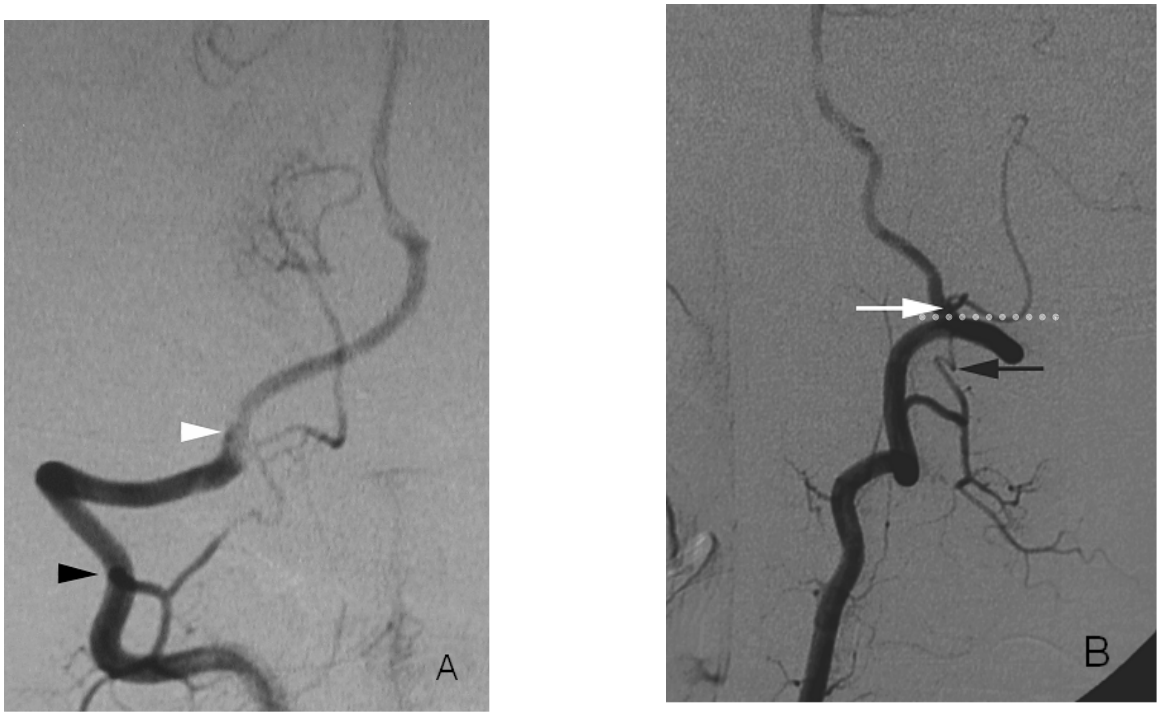


Fig. 15 (Case 5): A double proximal origin of the right PICA (LSA type) in 15-year-old boy.

A, DSA, right VA, anteroposterior view, showing a double origin of the right PICA from the right LSA via C1 (white arrowhead) and C2 (black arrowhead) roots. Note the location of the PICA origin from the inferior aspect of the VA, consistent with a variation of the LSA.

B, DSA, right VA, lateral view, showing the ascending course of the proximal PICA (white arrow) aligned with the LSA (black arrow). The proximal PICA has a more anterior position in the foramen magnum region (whose approximate anterior-posterior diameter is indicated by a dotted line, compare with Fig. 14B), a location consistent with the course of the LSA.

Case 6 – A 47-year-old woman presented with acute left lower quadranopsia and left-sided paresthesias after head trauma. Brain MR imaging revealed right cerebral peduncle and right occipital lobe strokes. Cerebral DSA showed a proximal right PCA dissection. Incidentally, a C1 origin of the left PICA was observed. This variant is linked to the PSA (Fig. 16 A and B).

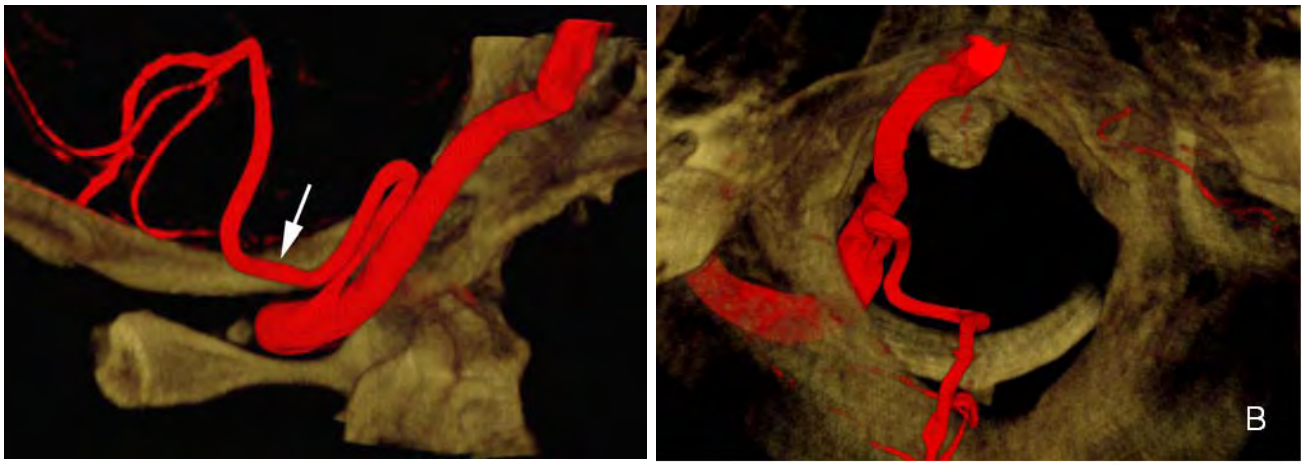


Fig. 16 (Case 6): A proximal (C1) origin of the left PICA (PSA type) in a 47-year-old woman, as demonstrated by a 3D reconstruction of a rotational angiogram with simultaneous osseous and vascular rendering. This technique is known as 3D fusion DSA (3D-FDSA) (Infinix, Toshiba, Japan).

A, 3D-FDSA, left VA, lateral view, showing that the proximal portion of the PICA follows the typical course of a PSA trunk as described by Maillot and Koritke (18), (i.e., a first segment closely paralleling the distal VA, followed by a sharp caudal and dorsal curve that brings the vessel in a posterior-lateral position within the foramen magnum). The ascending ramus of the PSA (white arrow) then continues as the proximal portion of the PICA.

B, 3D-FDSA, left VA, superior axial view, demonstrating the posterior location of the PICA within the foramen magnum. The topographical correspondence between the adult distal VA and PSA/PICA, and the embryonic ventral and dorsal radicular branches of C1 is well illustrated.

C2 origin of PICA

C2 origin of PICA

Case 7 – Cerebral DSA was obtained in a 44-year-old woman as part of investigations for transient ischemic attacks. The angiogram was unremarkable. Incidentally, a C2 origin of the right PICA was documented. This variant is linked to the PSA (Fig. 17 A and B).

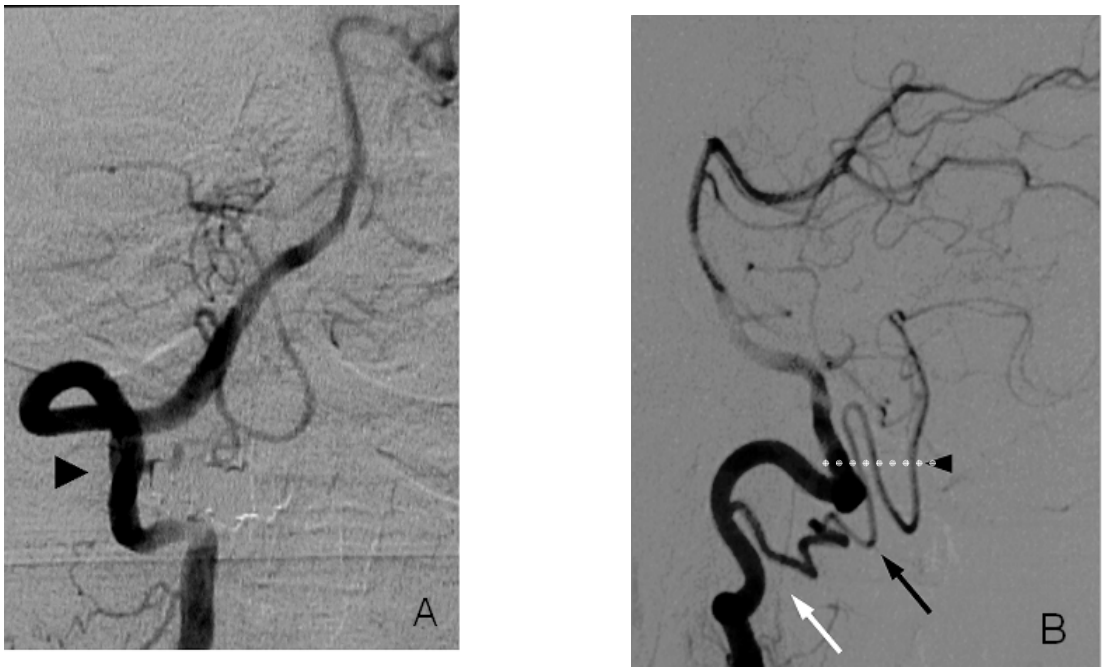


Fig. 17 (Case 7): A proximal (C2) origin of the right PICA (PSA type) in a 44-year-old woman.

A, DSA, right VA, anteroposterior view, showing the C2 origin of the right PICA (black arrowhead, partially masked by teeth subtraction artifacts).

B, DSA, right VA, lateral view, showing the posterior position of the PICA (black arrowhead) at the foramen magnum (whose approximate anterior-posterior diameter is indicated by a dotted line). The PICA is the cranial continuation of the posterior spinal axis (black arrow), fed by a segmental C2 branch. Note the more anterior position of the LSA (white arrow).

Case 8 – A 44-year-old woman was investigated for basal ganglia and subarachnoid hemorrhage. Cerebral DSA was unremarkable. Incidentally, a C2 origin of the PICA was noted on the left side. This variant is linked to the LSA (Fig. 18 A and B)

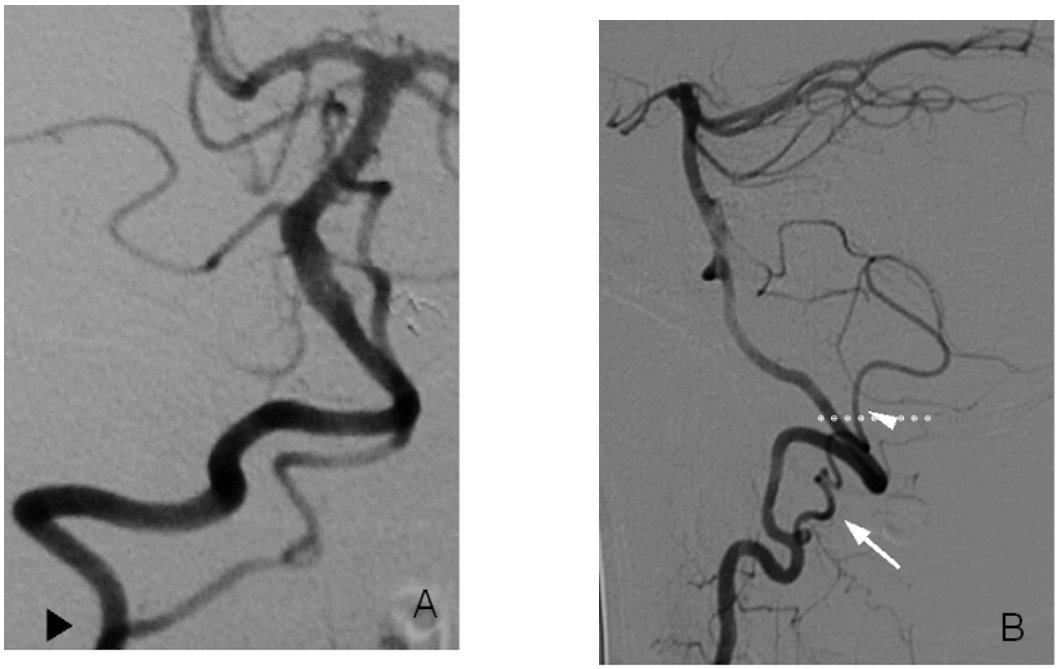


Fig. 18 (Case 8): A proximal (C2) origin of the left PICA (LSA type) in a 44-year-old woman (shown flipped horizontally for comparison with Fig. 17).

A, DSA, left VA, anteroposterior view, showing a proximal (C2) origin of the left PICA (black arrowhead).

B, DSA, left VA, lateral view, showing that the left PICA (white arrowhead) is the cranial continuation of the LSA (white arrow), with which it is aligned. Compared with Fig. 17B, the PICA crosses the foramen magnum (whose approximate anterior-posterior diameter is indicated by a dotted line) in a more anterior position consistent with the LSA rather than the PSA.

Duplication of the VA

Case 9 – A 47-year-old woman presented with acute left dysmetria. Brain MR imaging showed an acute stroke in the left cerebellar hemisphere. Cerebral DSA was normal. Incidentally, a duplication of the distal right VA was observed. This variant is linked to the PSA (Fig. 19A and B).

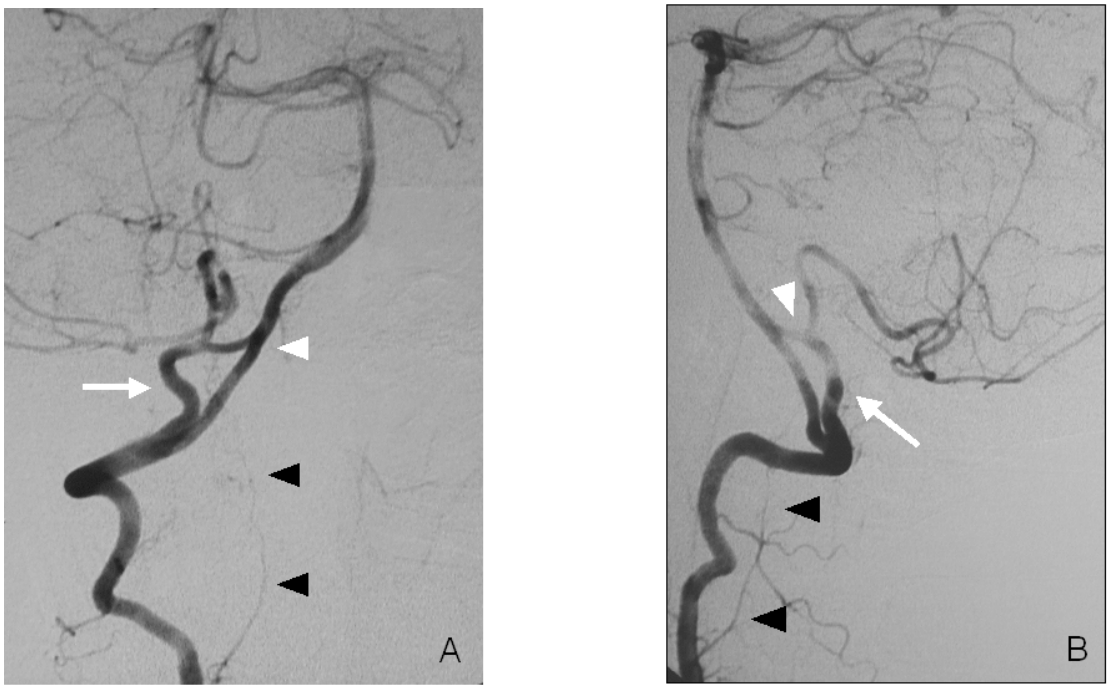


Fig.19 (Case 9): Duplication of the distal right VA (PSA type) in a 47-year-old woman.

A, DSA, right VA, anteroposterior view, showing a duplication of the distal VA consisting, in fact, of a double origin of the right PICA from the PSA (C1 origin, white arrow) and from the V4 segment of the VA (normal origin, white arrowhead). The LSA is indicated by black arrowheads.

B, DSA, right VA, lateral view, same also showing a duplication of the distal VA consisting of double origin of the right PICA from the PSA (C1 origin, white arrow) and from the V4 segment of the VA (normal origin, white arrowhead).

Case 10 – A 52–year-old woman was investigated for left pulsatile tinnitus. The angiogram was unremarkable. Incidentally, a duplication of the distal left VA was noted. This variant is linked to the LSA (Fig. 20 A and B).

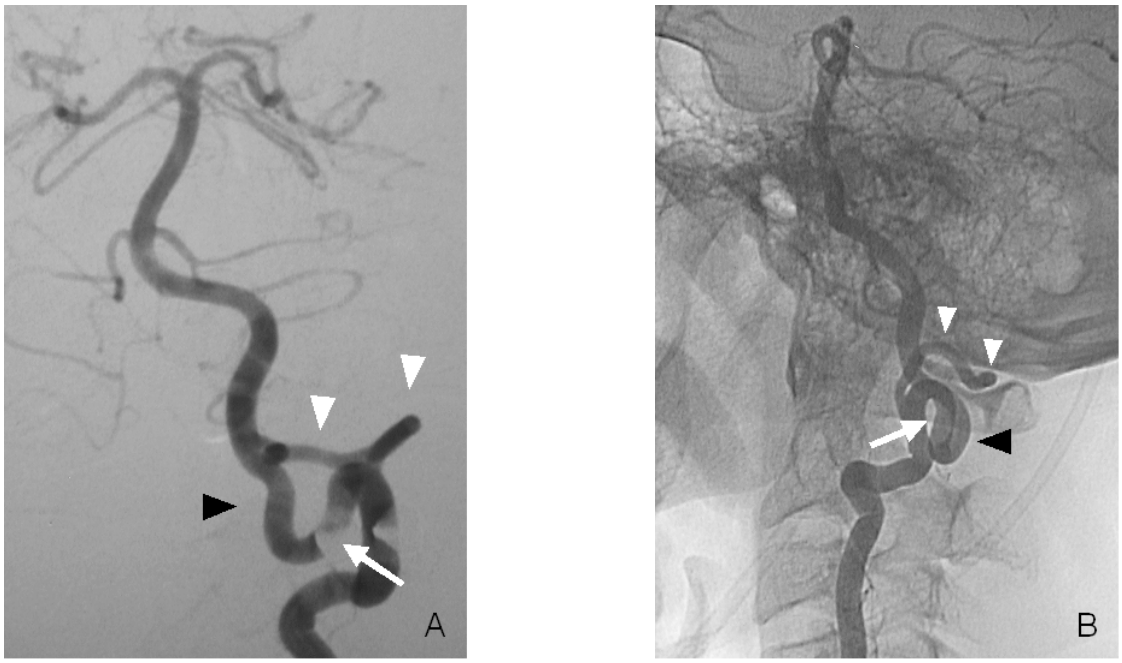


Fig. 20 (Case 10): Duplication of the distal left VA (LSA type) in a 52–year-old woman

A, DSA, left VA, anteroposterior view, showing a dominant intradural segment (black arrowhead) corresponding to the LSA with a prominent C2 root (white arrow). The normal distal VA is slightly hypoplastic (white arrowheads).

B, DSA, right VA, unsubtracted lateral view. As with Fig. 20A, this variant also shows a dominant intradural segment (black arrowhead) corresponding to the LSA with a prominent C2 root (white arrow).

Aberrant course of the distal VA

Case 11 – Brain MR imaging in a 64-year-old woman with systemic lupus erythematosus and progressive cognitive changes showed scattered foci of white matter T2-weighted hyperintensities suggesting cerebral vasculitis. Cerebral DSA was normal. Incidentally, a superior convexity of the distal right VA was observed. This variant is linked to the PSA (Fig. 21 A and B).

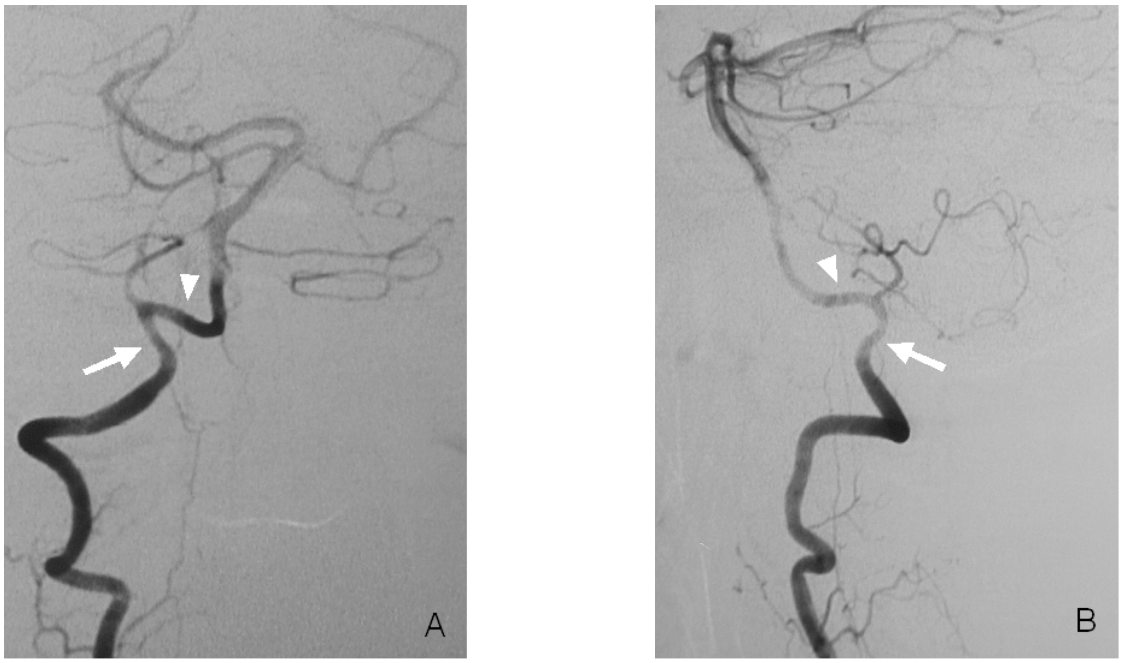


Fig. 21 (Case 11): Aberrant course of the distal right VA (PSA type) in a 64-year-old woman

A, DSA, right VA, anteroposterior view. This variant is similar to the VA duplication shown in Fig. 20, with a double origin of the PICA from the PSA (white arrow) and from the distal VA (white arrowhead). The difference between the two variants lies in the absence of a segment of the right VA between the two PICA origins, resulting in a seemingly “aberrant” course of the VA.

B, DSA, right VA, lateral view. As with Fig. 21A, this variant is also similar to the VA duplication shown in Fig. 20, with a double origin of the PICA from the PSA (white arrow) and from the distal VA (white arrowhead).

Case 12 – A 58-year-old woman presented with syncopal episodes suggesting vertebrobasilar insufficiency. Cerebral DSA was unremarkable. An intradural course of the right VA was noted (Fig. 22).

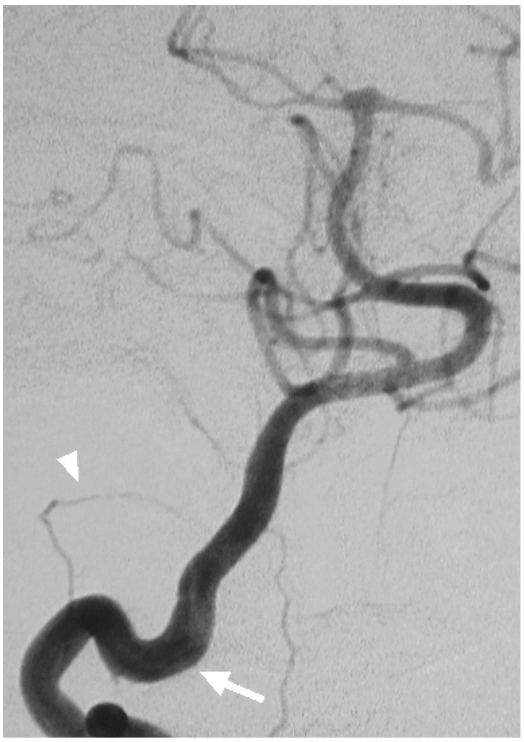


Fig. 22 (Case 12): An aberrant course of the distal right VA (LSA type) in a 58-year-old woman. DSA, right VA, anteroposterior view, showing the intradural course of the distal VA that is, in fact, a prominent segment of the LSA (white arrow). Note the presence of a very small vascular channel corresponding to the real distal VA (white arrowhead).

Discussion

C1 origin of PICA

A low PICA origin at the C1 level is a frequent variant, whose relation with the LSA was identified by Lasjaunias and co-authors (8). Rostrally, the LSA and the PICA are connected by small anastomoses located at the level of the restiform body. It is believed that, during embryonic development, one of these anastomoses remains prominent and establishes a longitudinal continuity between the PICA and the LSA, the PICA eventually originating from the extradural VA through the LSA. Case 5, which shows a double origin of the PICA from the VA at the C1 and C2 levels, illustrates this concept (Fig. 15). In this configuration, the proximal segment of the PICA corresponds to the LSA (Fig. 23E).

In Case 4, on the other hand, the PICA originates from the superior aspect of the distal VA, and adopts a “hairpin” course with a cranial loop before it passes posteriorly and ascends towards the cerebellum (Fig. 14). This typical hairpin course of the initial portion of the PICA strongly resembles the course of the radicular arteries joining the posterior longitudinal spinal axis. As mentioned above, the PSA embryologically corresponds to the descending ramus of the posterior radicular branch of C1, while the ascending ramus establishes small connections with the PICA. Indeed, small anastomoses between the PICA and the PSA have been described in the literature. Sabatier (118), Maillot and Koritke (18), and Lazorthes and colleagues (105), for instance, explicitly mentioned an ascending branch of the PSA that anastomoses with the PICA (Fig. 24). By analogy with Case 5, these anastomoses may be preferentially recruited during development to result in an apparent origin of the PICA from the PSA into the initial (Fig. 23B). Case 4 thus represents an alternative C1 origin of the PICA from the PSA system.

Case 6 shows another example of a low PICA origin at the C1 level from the PSA. Fig. 16 illustrates particularly well how the distal segment of the VA corresponds to the anterior radicular branch of C1 and how the proximal segment of the low PICA displays the features of the posterior radicular branch (PSA).

C2 origin of PICA

Case 7 shows a C2 origin of the PICA of the PSA type, and Case 8 the corresponding variant derived from the LSA system. The posterior location of the PICA at the level of the foramen magnum, the hairpin turn of its proximal segment, and the presence of a separate LSA help identifying the PSA-type variant. The LSA variant has, on the other hand, a C1-C2 course aligned with the expected course of the LSA, resulting in a more anterior position at the foramen magnum.

Duplication of the VA

Arterial duplication refers to the division of an artery into two distinct vessels with different courses. Duplications, in which a single arterial lumen divides into two parallel channels following the same course, are frequently confounded with fenestrations. Duplications of the VA derived from the LSA system have been described by Lasjaunias and co-authors. In this variation (Fig. 23F), the upper limb of the duplication corresponds to the VA *per se*, while the lower limb is a prominent intradural LSA segment (as in Case 10). Case 9, which shows a VA dividing into two vessels with separate courses, is also consistent with an arterial duplication. However, in this situation, the duplicated segment is more distal, and the supernumerary limb is located above rather that below the limb corresponding to the normal VA component (Fig. 23C). The upper limb of the duplication forms an upward loop, from which the PICA arises.

In this variant, we think that the upper limb results from the simultaneous persistence of both the normal origin of the PICA and of a second origin derived from the PSA system (equivalent to a low PICA origin, as shown in Case 4). Here again, the identification of a distinct LSA helps differentiating the two types of variations (*i.e.*, derived for the LSA or the PSA systems).

Aberrant course of the distal VA

Case 12 is an example of intradural course of the distal VA. In this variant, the apparent distal VA corresponds in fact to a prominent LSA, while the VA may be diminutive (as in our case) or absent. It is similar in nature to the VA duplication showed in Case 10, and can be viewed as a VA duplication in which the LSA limb is markedly dominant (Fig. 23F).

Case 11 shows a more distal type of “aberrant” course of the distal VA, where the vessel forms an upward loop that gives rise to the PICA. This type of aberrant course results from the simultaneous presence of two separate origins for the PICA, the normal one as well as a second origin derived from the PSA system. This situation is, in fact, similar to the distal VA duplication illustrated in Case 9, with the difference that the normal distal VA is, in this aberrant course variant, diminutive or absent (Fig. 23C).

It thus appears that a parallel can be drawn between duplication and aberrant course of the distal VA for variants derived from either the LSA and PSA systems. An aberrant course is an extreme form of duplication, in which the limb corresponding to the normal VA is diminutive or absent, while the “supernumerary” limb has become a markedly the dominant or unique vessel.

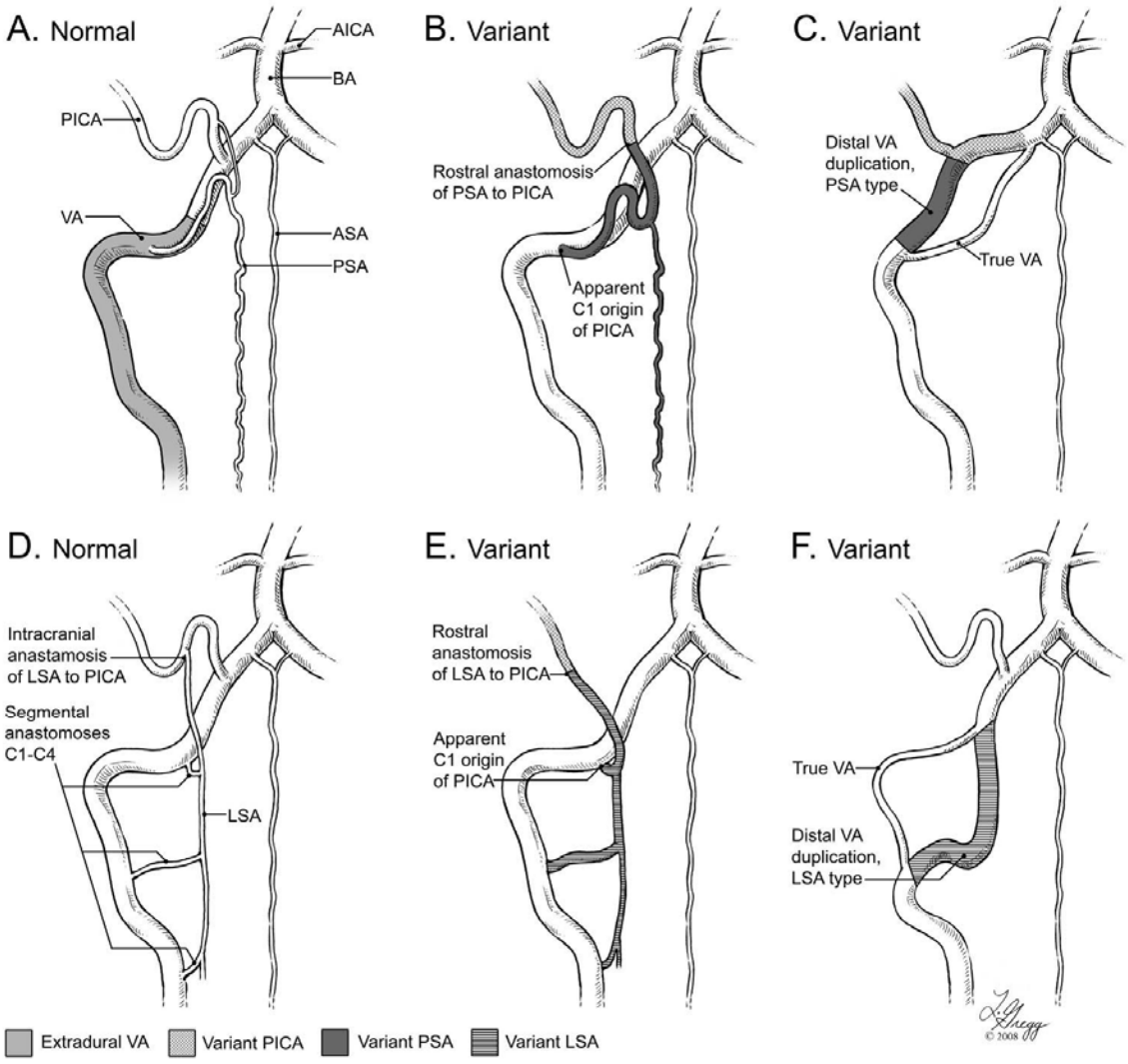


Fig. 23. Schematic representation of the normal anatomy of the PSA and LSA systems, as well as some of their variations (right side)

A, The extradural and intradural segments of the VA are shown in dark and light grey, respectively. The PSA originates from the extradural VA and parallels its course before bifurcating into a descending posterolateral ramus (the PSA per se), and an ascending ramus joining the PICA. The latter may become a variant origin of the PICA. The ASA is also shown.

B, The C1 origin of the PICA (PSA type). The anastomosis between the ascending ramus of the PSA and the PICA is prominent, and the PSA serves as the actual origin of the PICA.

C, A distal VA duplication of the PSA type, in which the true distal VA is paralleled by a supernumerary vascular segment that branches off the PICA. The relative size of the two limbs can vary. If the true distal VA is markedly diminutive or absent, the variant becomes an aberrant course of the VA (PSA type).

D, The LSA is the most lateral of the three cervical spinal axes. It establishes several segmental connections with the VA (C1-C4) as well as a cranial anastomosis with the PICA.

E, The C1,C2 and C3 origins of the PICA (LSA type). The rostral anastomosis of the LSA with the PICA is prominent, and any of the lateral segmental connections of the LSA with the VA may serve as a PICA origin, alone or in combination.

F, A duplication of the distal VA of the LSA type. This time, the true VA is cranial to the supernumerary limb, which is made of a prominent LSA segment. If the true distal VA is markedly diminutive or absent, the variant becomes an aberrant course of the VA (LSA type).

Mechanisms of formation

Why and in which circumstances these arterial variants forms is unknown. They may underlie specific hemodynamic contraits during embryogenesis or have a genetic background.

Clinical implications

The arterial variants presented in this section were discovered incidentally and can be considered as asymptomatic. Identifying low PICA origins from the vertebral artery might be of particular importance prior to cervical puncture (i.e.. for myelography)(119)or when planning spinal surgery in this region, in order to avoid hemorrhage. The portion of the PICA that is most likely damaged when using the C1-C2 puncture ist the caudal loop.

Conclusion

Anatomic variations commonly involve the distal segment of the VA. Lasjaunias and colleagues have simplified this complex anatomy by showing that a certain number of anatomic variants can be understood as segmental caliber variations of the distal VA and of one of its branches, the LSA (LSA). We show that a similar but not identical set of anatomic variants of the distal VA is linked to another one of its branches, the (PSA), as the result of comparable segmental alterations in caliber and connections.

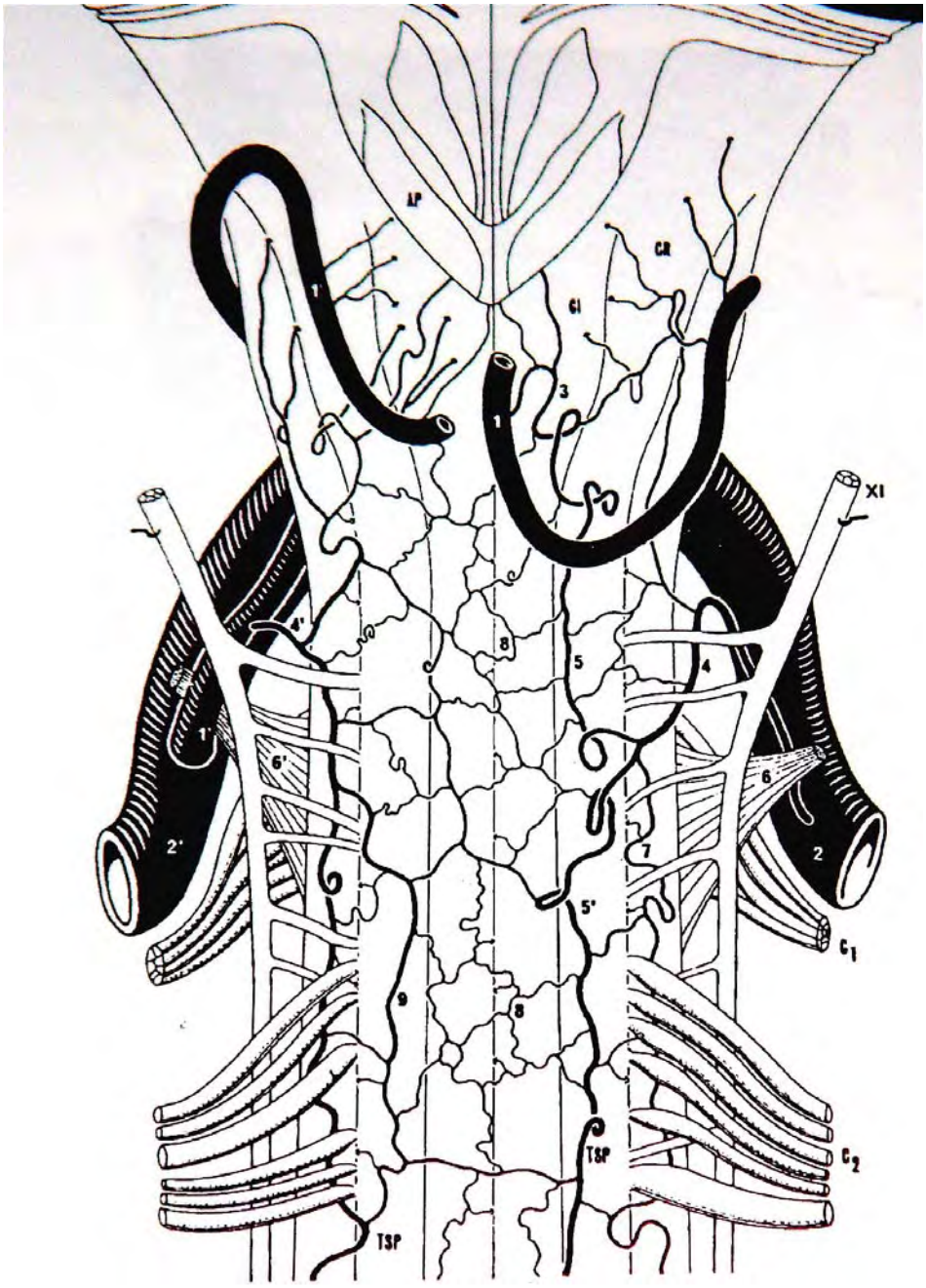


Fig. 24 (taken from Maillot and Koritke (18)): drawing illustrating the possible anastomoses between the PICA and the PSA (posterior spinal artery). 4 and 4' design PSA origins from the PICA and vertebral arteries respectively. Note the anastomosis between the PSA and the PICA (5), which, if recruited during embryological development and incorporated into the PICA, results in the PSA configuration of the PICA.

VARIATIONS IN THE VERTEBROBASILAR SYSTEM RELATED TO PARTIAL PERSISTENCE OF THE PRIMITIVE LATERAL BASILOVERTEBRAL ANASTOMOSIS OF PADGET

As outlined in the first part of this work, the PLBA develops as a cranial branch of the ProA and anastomoses with lateral branches of the developing basilar artery (6). Padget (4) anticipated that variations of the VA and its branches resulted from partial persistence of the PLBA. Moffat (6) recognized the LSA as extracranial equivalent of the PLBA. In the following section a series of anatomic variants of the vertebrobasilar system resulting from partial persistence of the PLBA are described and illustrated and their mechanism of formation is discussed.

Illustration of cases

Case 13 – A 55-year-old woman was investigated for suspicion of cerebral vasculitis. Digital subtraction angiography (DSA) was unremarkable. A left PTAV continuing as the left PICA was incidentally documented (Fig. 25). This “classic” PTAV can be understood as a primitive trigeminal artery originally connected to the PLBA rather than to the LNA. See also Fig. 34B.

Case 14 – A 64-year-old woman was investigated for suspicion of cerebral vasculitis. DSA was unremarkable. Incidentally, a right PTAV providing both the SCA and the AICA was documented (Fig. 26). This unusual PTAV with two cerebellar arteries can be understood as a “classic” PTAV with additional persistence of a segment of PLBA linking two cerebellar branches. See also Fig. 34C.

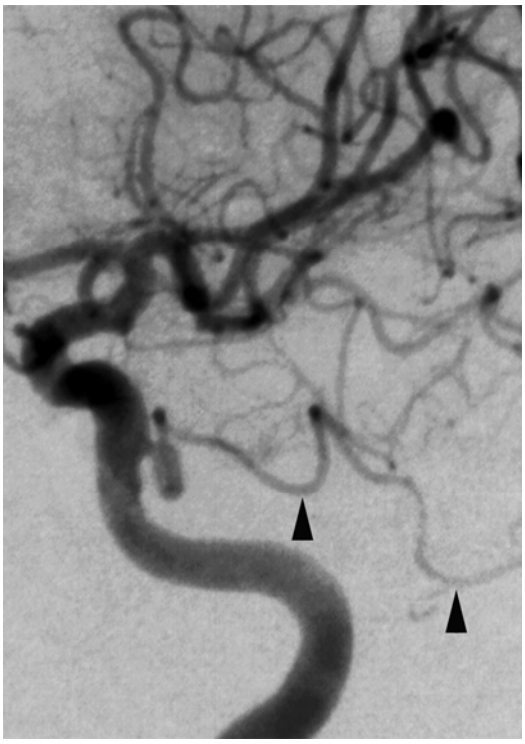


Fig. 25 (Case 13): 55-year-old woman with a left persistent trigeminal artery variant (PTAV). Digital subtraction angiography (DSA), left common carotid injection, left anterior oblique view. The PTAV originates from the cavernous segment of the left internal carotid artery and feeds the left posterior-inferior cerebellar artery (arrowheads). There is no opacification of the basilar artery.

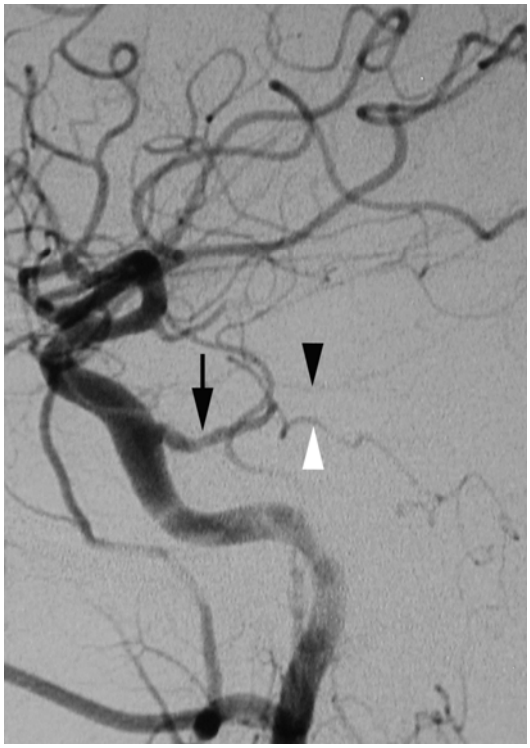


Fig. 26 (Case 14): 64-year-old woman with a right persistent trigeminal artery variant (PTAV). DSA, right common carotid injection, lateral view, showing a PTAV (arrow) providing both the superior cerebellar artery (black arrowheads) and the anterior inferior cerebellar artery (white arrowheads). There is no opacification of the basilar artery.

Case 15 – A 35-year-old woman underwent DSA after suspicion of intracranial aneurysm by MR angiography. DSA was unremarkable. A left PTAV connected both to the left AICA and the basilar artery was incidentally documented. This variant may be understood as a “classic” PTAV with an additional connection to the basilar artery. A true fetal configuration of the PCA was observed as well (Fig. 27). See also Fig. 34D.

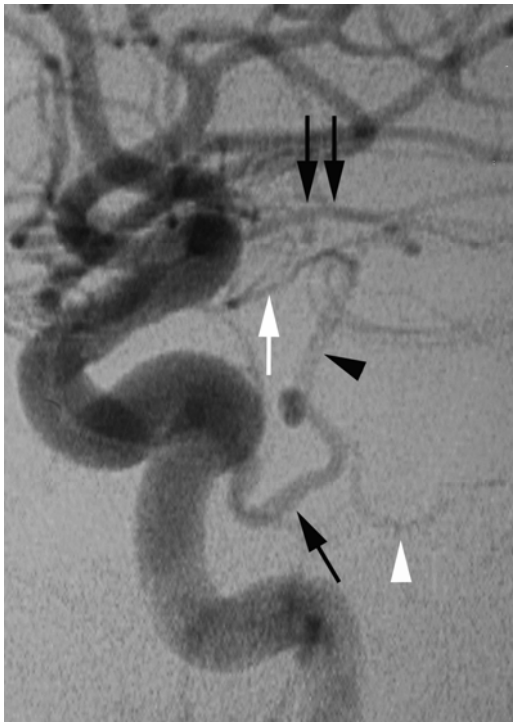


Fig. 27 (Case 15): 35-year-old woman with a left PTA/PTAV.

DSA, left common carotid injection, lateral view, showing a left PTAV (black arrow) that bifurcates to connect simultaneously to the basilar artery (black arrowhead) and to the left AICA (white arrowhead). This persistent anastomosis therefore shares the characteristics of a PTA and a PTAV. Also note the presence of a true fetal configuration of the left posterior cerebral artery, with the left posterior communicating artery ending into the basilar artery (white arrow), while the left anterior choroidal artery independently feeds the cortical territory of the posterior cerebral artery (double arrow).

Case 16 – A 44-year-old woman was investigated for subarachnoid hemorrhage. DSA showed a small VA aneurysm at the point of origin of a common trunk for the right AICA and PICA (Fig. 27A and B). This variant can be understood as a normal PICA with additional connection to the AICA via a partially persistent PLBA. See also Fig. 35A.

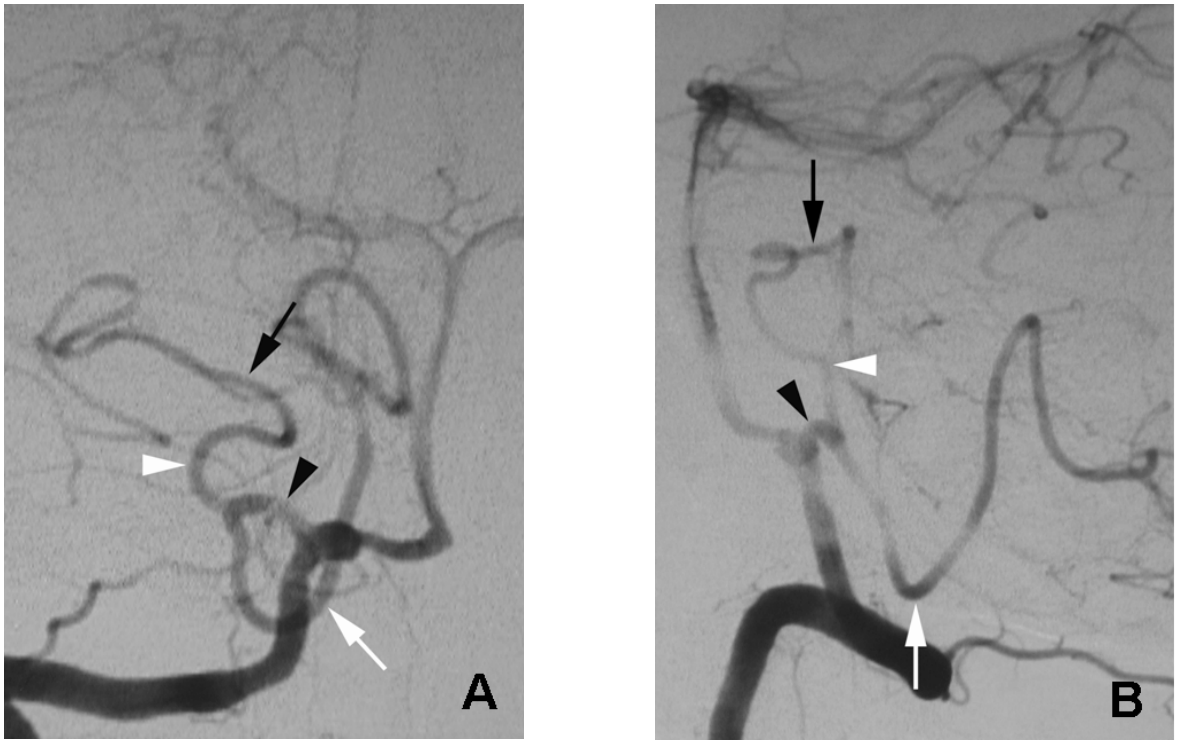


Fig. 28 (Case 16): 44-year-old woman with a common trunk for the right anterior-inferior cerebellar artery (AICA) and posterior-inferior cerebellar artery (PICA). **A,** DSA, right vertebral injection, transfacial view, showing the common trunk (black arrowhead) connecting to a persistent segment of primitive lateral basilovertebral anastomosis of Padget (PLBA) (white arrowhead). The persistent PLBA segment connects the right AICA (black arrow) to the right PICA (white arrow). **B,** DSA, right vertebral injection, lateral view, same legend as 27 A.

Case 17 – A 27-year-old man was investigated for subarachnoid hemorrhage. DSA showed a straight sinus thrombosis. Incidentally, the right AICA and PICA were seen originating from the right SCA via a long paramedian longitudinal channel (Fig. 29A and B). This variant can be understood as near-complete persistence of the right PLBA providing three cerebellar branches. See also Fig. 35 B.

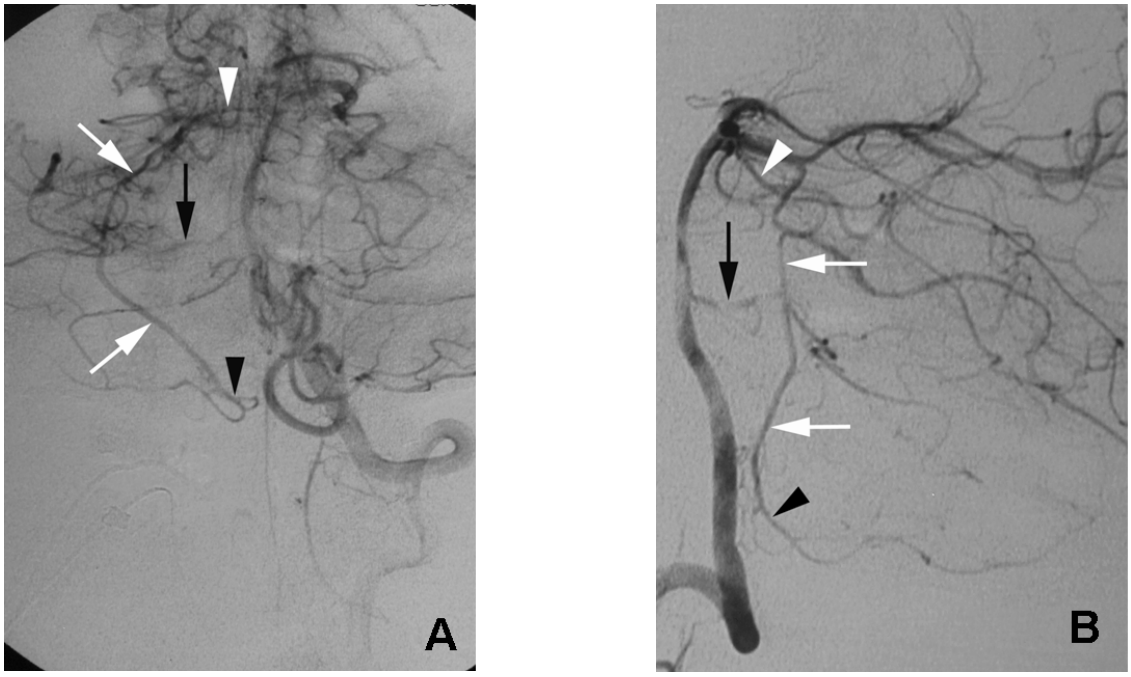


Fig. 29 (Case 17): 27-year-old man with persistent primitive lateral basilovertebral anastomosis of Padget (PLBA). **A**, DSA, left vertebral injection, transfacial view, showing partial persistence of the PLBA as a right paramedian longitudinal channel (white arrows) connecting the right superior cerebellar artery (white arrowhead) to the right anterior-inferior cerebellar artery (AICA) and posterior-inferior cerebellar artery (PICA) (black arrowhead). There is a connection between the PLBA and the basilar artery at the level of the right AICA (black arrow). **B**, DSA, right vertebral injection, lateral view, same legend as Fig. 29A. In addition, note a small plexiform connection between the caudal end of the PLBA and the V4 segment of the right vertebral artery. This connection topographically corresponds to the expected normal origin of the vertebral artery. The connection between the PLBA and the basilar artery (black arrow) at the level of the AICA is better opacified.

Case 18 – A 27-year-old woman was investigated for acute ischemic lesions involving both thalami, the mesencephalon, and the left occipital lobe. DSA was unremarkable, in particular the basilar artery was normal, and all the dural sinuses were patent. A vertebrobasilar duplication was incidentally documented (Fig. 30 A and B). This variant can be understood as partial proximal persistence of the right PLBA. See also Fig. 35 C.

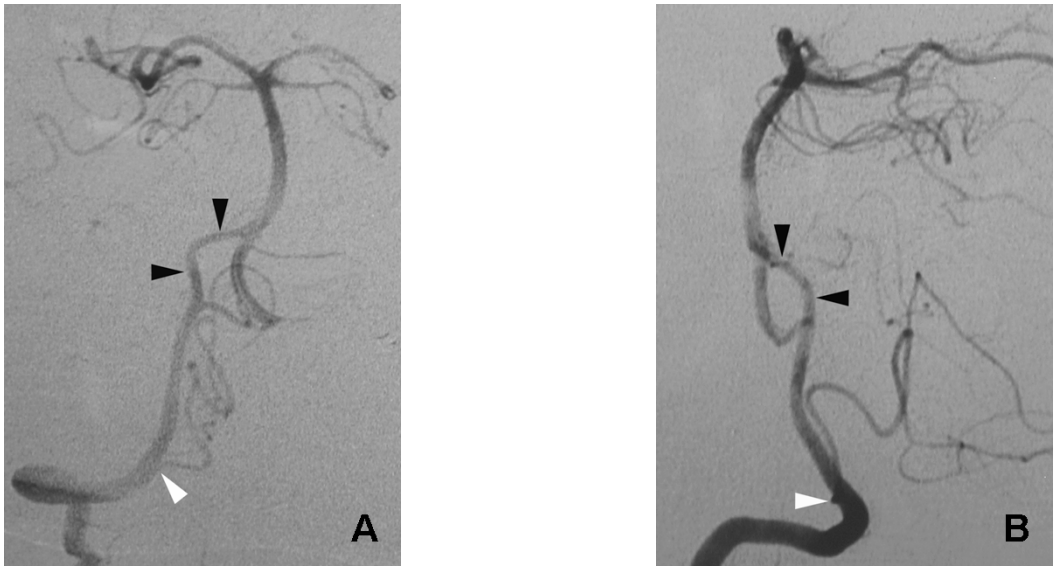


Fig. 30 (Case 18): 27-year-old woman partial vertebrobasilar duplication. **A,** DSA, right vertebral injection, transfacial view, showing an abnormal right paramedian channel (black arrowheads) connected caudally to the V4 segment of the vertebral artery (at the level of expected normal PICA origin), and cranially to the basilar artery (at the expected level of origin of the AICA). This channel corresponds to partially persistent right PLBA. An extradural origin of the PICA at the C1 level is seen as well (white arrowhead), which represents a variation of the posterior spinal axis (114). **B,** DSA, right vertebral injection, lateral view, same legend as 29A.

Case 19 – A 44-year-old woman was investigated for suspicion of cerebral vasculitis. DSA was consistent with bilateral vasculitis. Partial vertebrobasilar duplication, *i.e*., partial proximal persistence of the right PLBA, was incidentally documented (Fig. 31). See also Fig. 35 D.

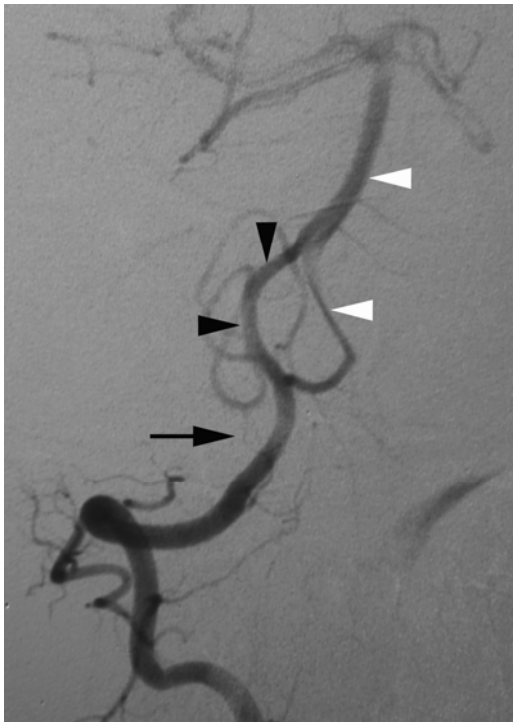


Fig. 31 (Case 19): 44-year-old woman with partial proximal persistence of the right PLBA. DSA, right vertebral injection, transfacial view, showing an abnormal right paramedian channel (black arrowheads) connected caudally to the V4 segment of the vertebral artery, and cranially to the basilar artery (white arrowheads). This variant is similar to the one documented in case 18, but shows in addition the origin of the right PICA from the persistent portion of the right PLBA. A small anastomosis between the PICA and the ascending ramus of the right posterior spinal artery can be seen (arrow). This normal anastomosis, when dominant, represents one of the mechanisms explaining an extradural origin of the PICA at C1 (114), like the one observed in Case18.

Case 20 – A 12-year-old girl with a suspicion of upper thoracic spinal vascular malformation by MR imaging. DSA confirmed the diagnosis of spinal AVM. Incidentally, partial vertebrobasilar duplication was documented (Fig. 32). See also Fig. 35 E.

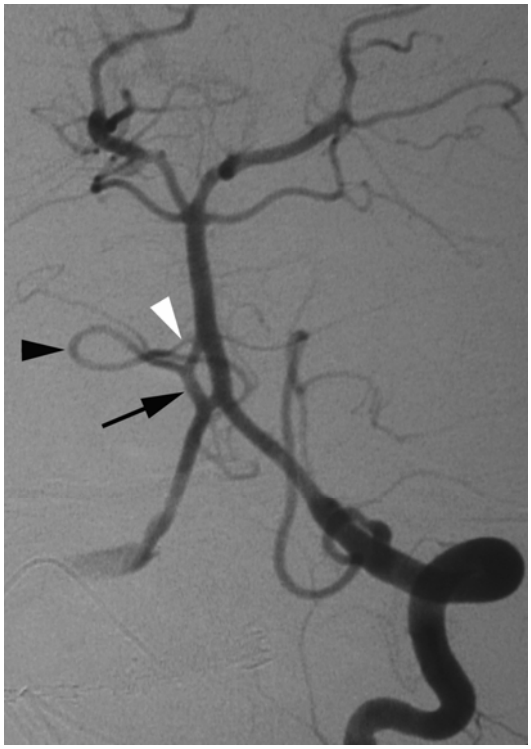


Fig. 32 (Case 20): 12-year-old girl with partial proximal persistence of the right PLBA. DSA, left vertebral injection, transfacial view, showing a right paramedian channel (black arrow) connected to the V4 segment of the vertebral artery, and to the basilar artery. In addition to the right PICA (black arrowhead), this persistent segment of the right PLBA also provides the right AICA (white arrowhead) immediately before its connection with the basilar artery. Note a C1 origin of the left PICA.

Case 21 – A 38-year-old woman with systemic lupus erythematosus and multiple cerebral infarcts was investigated for suspicion of vasculitis. DSA was unremarkable. Incidentally, a basilar origin of the right PICA and a left AICA-PICA trunk were observed (Fig. 33). See also Fig. 35F.

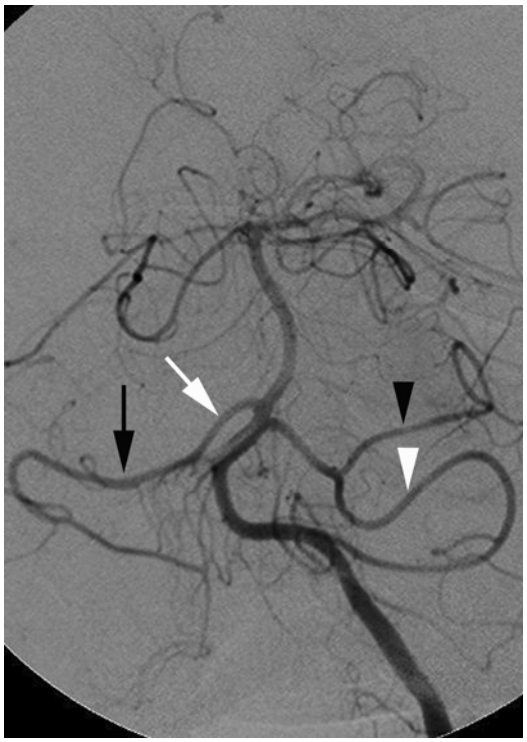


Fig. 33 (Case 21): 38-year-old woman with bilateral PICA origin variants. DSA, left vertebral injection, transfacial view, showing a basilar origin of the right PICA and a left AICA-PICA trunk. The basilar origin of the right PICA (black arrow) is a variation similar to cases 7 and 8, in which a persistent segment of PLBA (white arrow) results in partial vertebrobasilar duplication. In this case, however, a very small or absent proximal connection between the PLBA and the right vertebral artery results in an apparent basilar origin of the right PICA. The AICA-PICA trunk ("AICA-type") observed on the left side is a commonly encountered variation secondary to partial persistence of a PLBA segment linking the AICA (black arrowhead) to the PICA (white arrowhead).

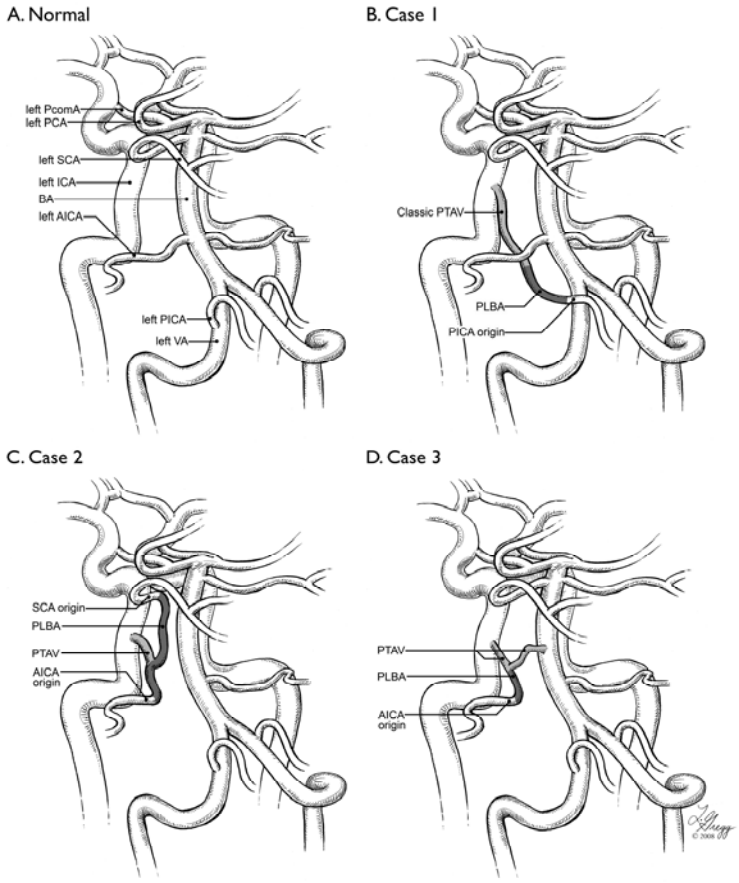


Fig. 35. Anatomy of typical and atypical persistent trigeminal artery variants (PTAV), left posterior oblique views. The variants are shown on the left side. Persistent PLBA segments are highlighted (light gray). **A**, Normal anatomy, with normal origin of the cerebellar arteries.

B, Typical PTAV. In this particular case, the left PICA originates from the cavernous segment of the left internal carotid artery. Any cerebellar artery can be involved in this type of variation. The drawing corresponds to Case 13. **C**, Atypical PTAV. In this configuration, a PTAV trunk provides more than one cerebellar artery. The drawing corresponds to Case 14, and shows a left PTAV branching off both the left SCA and AICA. The two cerebellar branches are connected by a segment of PLBA. **D**, Atypical PTA/PTAV. In this rare variation, the cavernous ICA trunk provides the left AICA (PTAV), but is also connected to the basilar artery (PTA). This drawing corresponds to Case 15. 1=posterior cerebral artery, 2=posterior communicating artery, 3=superior cerebellar artery (SCA), 4=anterior-inferior cerebellar artery (AICA), 5=posterior-inferior cerebellar artery (PICA)

Fig. 35. Anatomy of vertebrobasilar arterial variations related to the persistence of the primitive lateral basilovertebral anastomosis (PLBA) of Padget, ventral views. The PLBA is highlighted (light gray).

A, Right AICA-PICA trunk ("PICA-type"). The PICA remains connected to the right AICA via a segment of the PLBA that normally involutes. This drawing corresponds to Case 16.

B, Near complete persistence of the right PLBA. The persistent segment of PLBA connects the right SCA, AICA, and PICA. A small connection of the PLBA with the basilar artery is also shown; it corresponds to the transverse anastomosis that normally becomes the proximal portion of the AICA. Another small connection between the PLBA and the vertebral artery is shown, and represents the segment of PLBA that normally provides the proximal PICA. This drawing is similar to Case 17.

C, Vertebrobasilar duplication. The persistence of a segment of PLBA and of one of its transverse anastomoses creates a duplication of the vertebrobasilar junction. This is not a fenestration, as the two limbs truly represent independent arterial channels, or channels not intended to coalesce into a single artery (as opposed to the longitudinal neural arteries, which occasionally produce a basilar artery fenestration by fusing incompletely). In addition, an extracranial origin of the right PICA at C1 ("posterior spinal artery-type") is shown as well, completing the similarity with Case 18.

D, Vertebrobasilar duplication. This configuration is similar to the one in Fig. 35C, with the addition of the right PICA originating in this case from the duplication itself. This configuration, seen in Case 19, corresponds to a normal PICA connected to the basilar artery via a persistent segment of PLBA and a transverse basilar anastomosis. As shown later, this anatomy allows understanding how, through a different pattern of regression, the PICA can originate from the basilar artery rather than from the vertebral artery. Note a small connection between the right PICA and the ascending ramus of the posterior spinal artery. This anastomosis can be dominant and become the apparent origin of the PICA, as shown in Fig. 35C.

E, Vertebrobasilar duplication. In this case, the duplication provides both the right PICA and AICA. This drawing corresponds to Case 20.

F, Basilar origin of the right PICA and left AICA-PICA trunk. This drawing is similar to Case 21, with its bilateral variants. On the left, there is an AICA-PICA trunk of the "AICA-type". This variant can be put in relation with Fig. 35E and 35A in order to appreciate the full spectrum of AICA-PICA trunks, with dual supply (Fig. 35E, Case 20), single supply from the vertebral artery ("PICA-type", Fig 35A, Case 16), or single supply from the basilar artery ("AICA-type", Fig 35F, Case 21). Similarly, the basilar origin of the PICA shown on the right side can be paired with Fig. 35D to emphasize the regression patterns at play in the formation of a normal origin of the PICA from the vertebral artery (not illustrated), an origin of the PICA from the basilar artery (Fig. 35F, Case 21), or a dual PICA supply (Fig. 35D, Case 19).

Discussion

Cases 13 to 15 are examples of PTAVs. As discussed above, the trigeminal artery is a transient vessel that provides flow to the posterior circulation in early developmental stages. A PTA originates from the cavernous portion of the adult internal carotid artery and joins the mature basilar artery. When the PTA ends in one of the cerebellar arteries, it is referred to as a PTAV. Haughton and coworkers (120) postulated that PTAVs occur when the two LNAs fail to fuse, in which case an "accessory" basilar artery establishes the continuity between the PTA and the cerebellar artery. We believe these variants to be rather related to partial persistence of the PLBA.

Case 13 illustrates a "classic" PTAV supplying a SCA (Fig. 25). The PTAVs in cases 14 and 15 are somewhat unusual as they have dual terminations. In Case 14, the PTAV supplies both the AICA and the SCA through the persistence of a segment of the PLBA linking the two cerebellar arteries (Fig. 34C).

In Case 15, the PTAV is connected to the basilar artery as well as to the AICA (Fig. 34D). In this instance, the dual connection is due the persistence of the transverse connection established between the ipsilateral LNA and the PLBA (a connection that should normally become the proximal segment of the AICA) in addition to a connection established between the ICA and the PLBA. Although pre-segmental arteries normally join the LNA, Moffat has observed in the rat the occasional occurrence of a pre-segmental artery joining the PLBA, the latter keeping or not a small connection with the LNA (6) [3]. A "classic" PTAV occurs when this connection is absent, while a PTA/PTAV occurs when this connection is present. The fact that pre-segmental arteries generally join the LNA and rarely the PLBA should then explain why PTAs are more commonly seen than PTAVs. A variant comparable to our Case 15 (but labeled as an AICA originating from a PTA) was reported by Yamada *et al*. (121), while Arakawa and co-authors (122) made two similar observations during autopsies. Both groups of authors mentioned the role of the PLBA in the formation of these AICA variants. Fig. 34 shows how, in each case, a segment of the PLBA is incorporated in the PTAV.

Case 16 and Case 17 and 21 illustrate common trunks of origin for several cerebellar arteries. In Case 16, the trunk provides both the AICA and the PICA, while the same two branches originate from an AICA trunk in Case 21. In both cases the AICA and PICA are linked by a longitudinal arterial segment that corresponds to a partially persistent PLBA.

In Case 17, the common trunk provides the PICA, the AICA, and the SCA, the cerebellar arteries being connected by a paramedian longitudinal vessel that represents an almost complete persistence of the PLBA (Fig. 28 A and B). In Case 17, the common trunk provides the PICA, the AICA, and the SCA, the three cerebellar arteries being connected by a

[3] The pre-segmental artery that Moffat observed to be occasionally connected to the PLBA in the rat was the primitive otic artery. However, the primitive otic artery plays a dominant role in the rat comparable to the dominant role played by the primitive trigeminal artery in man.

longitudinal anastomosis that represents an almost complete persistence of the PLBA (Fig. 29 A and B). This persistent PLBA segment in Case 17 also has a transverse connection with the basilar artery at the level of the AICA, and a plexiform anastomosis with the V4 segment of the VA, at the level of the expected origin of a normal PICA. Except for the presence of a detectable continuity with the LSA, this anatomic configuration corresponds to the fetal appearance of the PLBA, *i.e.*, a paramedian channel connected medially to the vertebrobasilar axis and laterally to the three cerebellar arteries.

In cases 18 to 20, the vertebrobasilar junction is duplicated. This variant needs to be distinguished from a typical basilar artery fenestration, *i.e.*, from partial lack of fusion of the ascending rami of the anterior radicular branch of C1 if the fenestration is proximal, or partial lack of fusion of the LNA if the fenestration is distal. In a vertebrobasilar duplication, the lateral segment is due to the persistence of a portion of the PLBA, whereas the medial one corresponds to the usual course of the basilar artery. The role of the PLBA in the formation of vertebrobasilar duplications was initially proposed by De Caro and co-authors in 1991 (123). A second observation was published by Yagi et al. in 2004 (124). We offer three additional examples of this variation, each illustrating a different pattern of cerebellar branching from the persistent PLBA (no cerebellar branch in Case 18, one in Case 19, two in Case 20). In addition, a C1 origin of the PICA (PSA type) is seen in cases 18 and 20, while a small anastomosis between the right PSA and the PICA is seen in Case 19. Low PICA origins at the C1 level represent variants of either the PSA or the LSA systems, both types having distinct angiographic characteristics (114). The low PICAs in cases 18 and 20 are of the PSA type. The small anastomosis seen in Case 19 illustrates the normal connection that has become abnormally dominant in cases 18 and 20, resulting in the apparent origin of the PICA from the PSA.

In cases 19 and 20, the ipsilateral PICA originates from the lateral segment of the vertebrobasilar duplication, a finding consistent with the hypothesis that the normal intradural origin of the PICA is derived from the proximal portion of the PLBA. This type of variant exemplifies how the PICA can originate either from the V4 segment of the VA ("normal origin") or from the proximal basilar artery, depending on which portion of the PLBA becomes dominant at the adult stage. A basilar origin of the PICA is shown in Case 21 (right side). In Case 20, the persistent PLBA branches off both a PICA and an AICA before connecting to the basilar artery. This variant allows understanding how an AICA-PICA trunk, a relatively common variant, can originate either from the VA ("PICA-type") or from the basilar artery ("AICA-type"), depending on which part of the PLBA become dominant. As mentioned above, Case 16 shows an AICA-PICA trunk of the "PICA-type", while Case 21 (left side) demonstrates an AICA-PICA trunk of the "AICA-type".

Of note, if the persistence of a significant portion of the PLBA is rare in man, it can be observed with various frequencies in other species. For example, the PLBA persists in its entirety at the adult stage in the rat (6), while a PLBA-derived vertebrobasilar duplication is often observed in the dog (although not specifically labeled as such by the authors) (125).

Mechanisms of formation

The etiology of these arterial variations is unknown. As mentioned in the previous section, they might underlie specific hemodynamic contraits during embryogenesis or have a genetic background.

Clinical implications

Apart from one case, variants related to the PLBA were asymptomatic and found incidentally. In case 16, a small aneurysm was seen at the tip of a common AICA-PICA trunk. It is

possible that the sharp bifurcation of the common AICA-PICA trunk creates hemodynamically favorable conditions for the development of aneurysms, which might be true for similar variations. 3 other cases of aneurysms associated with an AICA-PICA trunk have been described (126-128). For clinical purposes, it should be kept in mind that in case of ischemia of a common trunk for 2 or more cerebellar arteries (as presented in cases 16, 17 and 21), lesions in the territories of more than one cerebellar artery may occur. Variations involving a common AICA-PICA trunk can be associated with compression of the facial nerve causing hemifacial spasm (129). Knowledge of the existence of an associated PTA or PTAV is important because of the possibility of embolization from carotid lesions to the posterior circulation. This is particularly relevant before surgical interventions in the carotid arterial system or during parasellar surgery, in order to avoid embolization and massive hemorrhage, or when planning endovascular interventions or procedures, such as a Wada test or a carotid artery balloon occlusion test (98).

Conclusion

Padget (4) described a transient longitudinal anastomosis between the developing vertebral and basilar arteries in the human embryo, which she named the PLBA. She anticipated that remnants of this vessel could result in anatomic variations of the VA and its branches. This report confirms Padget hypothesis by illustrating several unusual variants of the vertebrobasilar system derived from the PLBA, including a case of near-complete PLBA persistence, a PTA/PTAV combination, a PTAV trunk for two cerebellar arteries, and several examples of a rare type of vertebrobasilar duplication. In addition, partial persistence of the PLBA also explains the formation of more commonly observed variations, such as a classic PTAV or a classic AICA-PICA trunk.

NONINVASIVE ANGIOGRAPHY

We mainly used digital subtraction angiography (DSA) to illustrate the clinical variations presented in this work, as this technique constitutes the gold standard in neurovascular imaging. However, DSA carries a risk of stroke of 1.3% (130) and relies on skilled and experienced angiographers. In recent years, noninvasive imaging techniques such as CT and MR angiography (CTA and MRA respectively) have assumed a considerable role in this field.

MRA is a noninvasive magnetic resonance technique that can be applied to the intracranial or extracranial vasculature. In clinical settings, most frequently time-of-flight (TOF) and contrast material-enhanced MRA are used for this purpose. Apart from being noninvasive, MRA has the advantage of offering an excellent concomitant cerebral tissue visualization without ionizing radiation. Disadvantages include technical limitations in determining the exact relationship between vessels and osseous anatomy or aerated structures and, with TOF-MRA, in evaluating vascular stenosis or aneurysms (because of loss of flow-related intravascular signal intensity or inhomogenous signal intensity). MRA is contraindicated in carriers of electrical and electromagnetic devices (pacemakers) and of ferromagnetic material.

CTA is a fast, thin section, volumetric spiral CT examination performed with a time-optimized bolus of contrast material for the opacification of vessels. Compared to MRA it offers the advantage of being more readily available and less expensive, and having a close to 100% sensitivity for hemorrhage. Comparative studies suggest that overall, CTA may be slightly superior to MRA for the diagnosis of aneurysms and intra- as well as extracranial stenotic disease (131). However, it offers a less precise visualization of the brain parenchyma and exposes the patient to ionizing radiation.

Overall, DSA remains the gold standard in neurovascular imaging and should always be carried out when facing unclear diagnostic issues (i.e. when noninvasive imaging techniques leave doubt about the true nature of vascular lesions), this is even more important when a therapeutic consequence is derived from the diagnostic result.

LIST OF ABBREVIATIONS

AICA	**anterior inferior cerebellar artery**
ASA	**anterior spinal artery**
LSA	**lateral spinal artery**
LNA	**longitudinal neural arteries**
PCA	**posterior cerebral artery**
PComA	**posterior communicating artery**
PICA	**posterior inferior cerebellar artery**
PLBA	**primitive lateral basilovertebral anastomosis of Padget**
ProA	**proatlantal artery**
PSA	**posterior spinal artery**
PTA	**persistent trigeminal artery**
PTAV	**persistent trigeminal artery variant**
SCA	**superior cerebellar artery**

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