Pathology of the CEREBRAL BLOOD VESSELS

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With 322 illustrations

The C. V. Mosby Company

Saint Louis 1972

Printed in the United States of America

International Standard Book Number 0-8016-4767-3

Library of Congress Catalog Card Number 70-185526

Distributed in Great Britain by Henry Kimpton, London

PREFACE

The high mortality and frequent disability occasioned by diseases of blood vessels supplying the brain have engendered much disquiet. Notwithstanding the fact that vital statistics indicate cerebrovascular diseases to be of unquestionably greater import than nonvascular diseases of the brain, current textbooks of general pathology and neuropathology deal only superficially with cerebral blood vessels. Although the clinical aspects have not been neglected, pathology has assumed but a minor role. This state of affairs is a reflection of the perfunctory interest in cerebrovascular diseases displayed by neuropathologists and general pathologists, and so it is not surprising that the field has not been investigated intensively. Advances in electron microscopy, histochemistry, physiology, and pathology have made apparent the serious need for a text dealing exclusively with the cerebral blood vessels, which differ structurally, physiologically, and pathologically from those of extracranial sites. This, therefore, is a comprehensive treatise on diseases of the blood vessels supplying the brain and spinal cord. The coverage is not encyclopedic, being restricted of necessity by publication costs. Historical aspects have been for the most part omitted and comparative pathology touched upon lightly. Nevertheless, the mass of information widely dispersed in the literature has been reviewed and appraisals of the conflicting theories so rife in the field are proffered, with emphasis on the etiology and underlying mechanisms in the pathogenesis of disease. The application of new information and concepts in general pathology and physiology, both experimental and observational, adds considerably to the scope of the book. The bibliography is extensive. The book is intended for general pathologists, neuropathologists, neurologists, neurosurgeons, and neuroradiologists and as a reference book for psychiatrists, students, and clinicians in general.

It is my earnest desire that criticisms and provocative speculations will not only focus more attention on cerebrovascular diseases in general, but will stimulate a renewal of interest in scientific research and investigations in this field, which has been so long neglected.

Sincere thanks are due to Dr. George Margolis, Dr. V. J. McGovern, Professor F. R. Magarey, Dr. K. D. Barron, and Dr. V. N. Tompkins for making material available and to the authors acknowledged in the text who have permitted citations of their work or reproductions of their illustrations. The librarians of the Albany

Medical College and Albany Veterans Administration Hospital are especially remembered for their patience and invaluable assistance.

My thanks are also due to Miss B. McLean, Mrs. M. Egy, Miss S. Ingvarsson, and Mr. J. E. Walden, who assisted in various ways during the preparation of the manuscript and to the Department of Medical Illustration of the Veterans Administration Hospital for some of the photography. Dr. W. A. Thomas and Dr. K. D. Barron read the manuscript.

The investigations incorporated in the text were supported financially by the National Heart Foundation of Australia, N.I.H. Grants HE 12920 and HE 14177, and Veterans Administration Research Funds.

I am indebted to my mother for providing the opportunity to study medicine and I acknowledge with gratitude the continued encouragement, aid, and interest of my wife, Jean.

William E. Stehbens

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1 / Anatomy of the blood vessels of the brain and spinal cord

The vascular system of the brain and spinal cord exhibits morphological peculiarities not seen in other organs. The reason for the differences appears to be bound up with phenomena responsible for the unique functions of the highly specialized tissues of the central nervous system.

Arterial supply to the brain

The arterial supply to the brain is derived from two carotid and two vertebral arteries, which enter the skull through its base.

Common carotid artery

Anatomy. The right common carotid artery begins at the bifurcation of the innominate (brachiocephalic)[221] artery. The left arises independently from the aortic arch. Each of these arteries terminates by dividing into internal and external carotid arteries at the level of the upper border of the thyroid cartilage in the environs of the fourth cervical vertebra.

Variations. The point of origin of the three great vessels from the arch varies[58] as does the number of vessels arising from it. A "normal" pattern is found in 65% to 70%.[124] The right common carotid artery can arise independently from the aorta and its site of origin is inconstant.[27] The left common carotid artery is even more variable than the right, not infrequently arising from a common stem with the right common carotid and subclavian arteries. This was observed nine times in 130 subjects.[221] In rare instances, the common carotid arteries are absent and then the internal and external carotid arteries arise directly from the aorta.[7]

The termination of the common carotid may be as high as C1 or as low as T2.[124] Quain,[173] investigating the level of bifurcation in 295 subjects, found the bifurcation at the normal level in 184 individuals (62.4%), at the level of the hyoid bone in 60 subjects (29.4%), above the hyoid bone in 10 subjects (3.4%), at the middle of the thyroid cartilage in 26 subjects (8.8%), and opposite the cricoid cartilage in five subjects (1.6%).

Toole and Patel[223] found the bifurcation at the "normal" level in only 50% of individuals and Lie[124] ascertained it was usually at a higher level in children than in adults. Periodically a common carotid artery will not divide at all, but is continued directly into the internal carotid artery whence branches, generally arising from the external carotid artery, take origin.

Internal carotid artery

Anatomy. The internal carotid artery may conveniently be divided into four segments: cervical, petrosal, cavernous, and terminal or cerebral. The cervical segment lies posterolateral to the commencement of the external carotid. The initial portion and the terminal section of the common carotid form a slight fusiform dilatation, the carotid sinus.

The cervical segment of the internal carotid artery ascends in the neck veering with some tortuosity laterally and dorsally to the base of the skull. The artery then enters the carotid canal in the petrous portion of the temporal bone where it is closely related to the eustachian tube and tympanum.[198] This then, the petrous segment, courses upward and bends abruptly, passing almost horizontally forward and medially to the apex of the petrous temporal bone. It leaves the canal, traversing

the upper part of the foramen lacerum, ascends along the side of the body of the sphenoid bone, and enters the middle cranial fossa. It is extradural in position for a short distance and surrounded by areolar tissue before entering the cavernous sinus where angiograms show a circular constriction.[221]

The cavernous segment ascends for a short distance and proceeds horizontally forward until it underlies the anterior clinoid process. It then bends upward, grooving the medial side of the anterior clinoid process to leave the sinus. If both anterior and middle clinoid processes are bridged by bone, it leaves the sinus through the foramen thus formed. The carotid groove is an S-shaped shallow indentation on the side of the sphenoid body, where the artery lies close to the sphenoid sinus.[198] While in the cavernous sinus, the artery is supported by numerous fibrous strands and trabeculae that give the sinus a coarse spongelike appearance yet is excluded from the circulation by the sinus endothelium. On its lateral side, from above down, lie the oculomotor, trochlear, and ophthalmic division of the trigeminal and the abducent nerves. The carotid artery traverses the dura mater, and its terminal or cerebral segment (somewhat narrowed in angiograms) bends backwards below the optic nerve, from whence it travels upward and laterally, with the optic nerve and chiasm lying medially and the oculomotor nerve laterally. Below the anterior perforated substance it divides terminally into the middle and anterior cerebral arteries. The cerebral segment is supraclinoid in position and the very variable direction is best appreciated angiographically. The carotid siphon is the series of curvatures made by the cavernous and cerebral segments of the internal carotid artery. Angiograms demonstrate the tortuosity well (Fig. 1-1). The internal carotid artery does not branch in the neck.* The small caroticotympanic artery arises from the petrous segment in the carotid canal and enters the tympanum through a small canal in the petrous temporal bone. An inconstant pterygoid branch passes to the pterygoid canal to anastomose with the artery of the canal. Several small branches arise from the cavernous segment

*Branches of the external carotid artery occasionally arise from the internal or common carotid artery instead.[151]

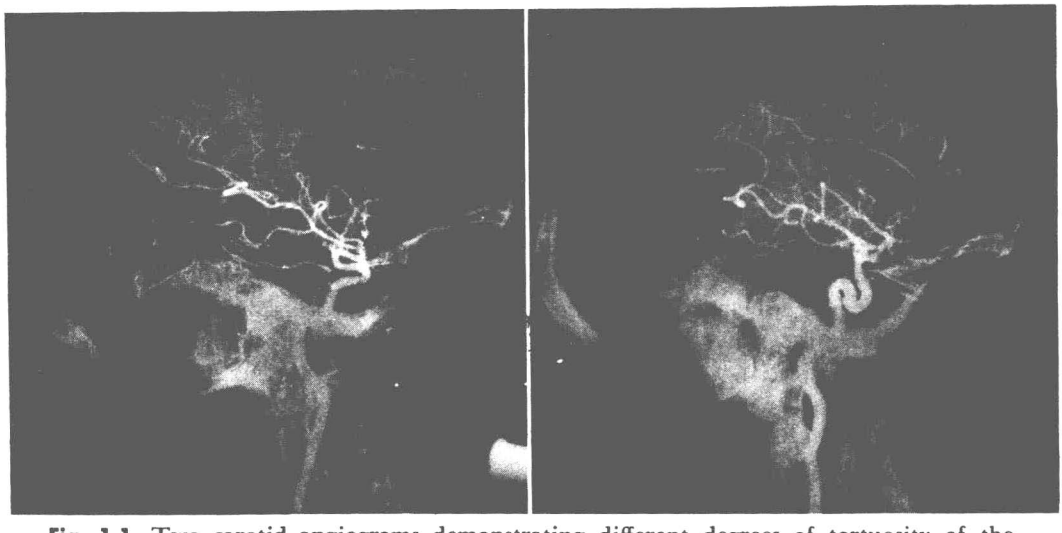

Fig. 1-1. Two carotid angiograms demonstrating different degrees of tortuosity of the siphon. Terminal segments of internal carotid arteries are smaller than the more proximal segments despite absence of branches of significant size.

and disperse to the nerves within the sinus, the walls of the cavernous and petrosal sinuses and the dura mater of the anterior and posterior fossae. Parkinson[162] described the following:

1. A meningohypophyseal trunk (constant in 200 cases) sends branches to the tentorium, the clivus and dorsum sellae, and hypophysis cerebri
2. An artery supplying the walls of the inferior cavernous sinus (present in 80% of the dissections)
3. A capsular artery (an intercarotid anastomosis anterior to the hypophysis)
4. An anterior meningeal branch assisting in the supply of the dura in the anterior cranial fossa[221]

Two additional branches of the cavernous segment, the marginal tentorial artery and the basal tentorial artery, arise at the same level as the primitive trigeminal artery[124] and supply the tentorium cerebelli. The marginal tentorial artery is of importance angiographically when associated with (1) increased intracranial pressure caused by supratentorial lesions and (2) increased vascularity caused by arteriovenous formations, ependymoma, acoustic nerve tumor, and malignant gliomas[204] and meningiomas of the tentorium[124] and falx.[20,67] The superior hypophyseal artery, a perforating branch from the internal carotid near its bifurcation, proceeds to the tuber cinereum, the pituitary stalk, and the neurohypophysis. Blood supply to the adenohypophysis is from a complex portal system extending from the tuber cinereum via the stalk to the adenohypophysis. Collateral vessels may be found between this portal system and neighboring hypothalamic vessels.[105,172]

All major branches of the internal carotid artery arise from the terminal or cerebral segment, that is, the ophthalmic, posterior communicating, anterior choroidal, anterior cerebral, and middle cerebral arteries.

Variations

Level of origin. The level at which the internal carotid commences is variable and the artery appears to undergo disproportionate lengthening during postnatal growth.

Tortuosity, coiling, and kinking of the cervical segment. The cervical segment often exhibits tortuosity, at times bilateral. Such sigmoid tortuosity increases the risk of injury to the carotid artery at tonsillectomy and indeed, on occasions, presents as a pulsating swelling bulging into the pharynx.[108,109]

Cairney[35] dissected the carotid vessels in 36 elderly subjects and found definite tortuosity, bilateral in two instances, in 12 of 76 arteries. The first bend was medially or otherwise laterally directed with variation also in the plane of the tortuosity. In 20 fetuses (5 months' gestation to full term), there was tortuosity in 4 (bilateral in one instance). Parkinson and associates[163] described 40 patients with a pulsatile swelling in the neck caused by a kink in the common carotid artery. They stressed the important role of hypertension, arteriosclerosis, and hemodynamics in the etiology. Severe degrees of tortuosity affect the internal carotid more often than the common carotid artery and angiography reveals the frequent nature of such changes.

Quattlebaum and associates[174] reported three elderly patients with transient hemiparesis. Pronounced elongation with kinking was accentuated in certain positions of the head. In 1000 consecutive angiograms Metz and associates[140] found kinking (angulation of 90% or less) or the formation of a complete loop in 161 cases (16%). They occur in every decade but predominantly between 41 and 70 years with mean age and blood pressure in the more severe degrees of kinking less than in subjects with milder grades of tortuosity. In another series[15] of 71 subjects diagnosed clinically as occlusive cerebrovascular disease, tortuosity or kinking of the carotid arteries was present in 17 individuals (24%) mostly with advanced atherosclerosis.

Weibel and Fields[237] investigating such arterial configurations divided them into three groups:

Table 1-1. Prevalence of carotid tortuosity in angiograms*

Angiography	Age group	Bilateral tortuosity	Unilateral tortuosity	Bilateral coiling	Unilateral coiling	Bilateral kinking	Unilateral kinking
Bilateral	Under 50 years	15%	6%	3%	3%	0.5%	3%
	Over 50 years	25%	12%	4%	3%	2%	4%
Unilateral	Under 50 years		27%		2%		4%
	Over 50 years		59%		4%		3%

*From Weibel, J., and Fields, W. S.: Tortuosity, coiling, and kinking of the internal carotid artery. 1. Etiology and radiographic anatomy, Neurology **15**:7, 1965.

1. *Tortuosity.* Any S- or C-shaped elongation or undulation in the course of the internal carotid artery
2. *Coiling.* Elongation or redundancy of the internal carotid artery resulting in an exaggerated S-shaped curve or in a circular configuration
3. *Kinking.* Angulation of one or more segments of the internal carotid artery associated with stenosis in the affected segment

The need for classification of the degree of tortuosity and redundancy of the artery is obvious but the above classification is not ideal. Weibel and Fields[237] examined 2453 carotid arteries from 1407 subjects. The proximal end of the tortuous segment of the internal carotid artery was seen most frequently at from 2 to 8 cm. above the common carotid bifurcation (peak incidence at about 3 cm.), coiling occurred most often between 4 and 9 cm. (peak incidence around 6 cm.), while kinking occurred mostly between 2 and 5 cm. (peak at about 3 cm.). In each case there was wide variation and bilateral symmetry was frequent. The prevalence of tortuosity, coiling, and kinking is shown in Table 1-1. Tortuosity is more frequent in older patients and kinking is associated with tortuosity more often than with coiling of the internal carotid arteries. Of the group having bilateral angiograms, 29% of those with bilateral tortuosity had neither a stenotic lesion nor an aneurysm as against 37% of those with unilateral tortuosity. This incidence of obviously severe degenerative changes whether primary or secondary is inordinately high. Those who were submitted to unilateral angiography only, included many young patients. Of this group, 65% with tortuosity had neither stenosis nor aneurysms compared to 76% in the group displaying no tortuosity, coiling, or kinking.

Despite (1) the greatest prevalence of tortuosity in the middle aged and elderly and (2) the greater prevalence of aneurysms and atherosclerotic stenotic lesions in association with tortuosity, Weibel and Fields[237] supported the contention[108,109] that tortuosity is caused by the persistence of a curvature at the junction of the third aortic arch and dorsal aorta (fifth week of embryonic life) from which vessels the internal carotid develops. The main support for this concept appears to be (1) the occurrence of tortuosity among some of the younger patients investigated by them, (2) the finding of tortuosity in some fetuses by Cairney,[35] and (3) the 12.5% incidence of tortuosity or coiling in 70 carotid angiograms of children under 15 years by Lie.[124] However bilateral tortuosity can hardly be regarded as supporting a developmental origin, for the same findings can be used equally well to substantiate a theory of widespread degenerative change in arteries even of the young. Fisher[60] found that pathological examination of histological sections from two specimens revealed comparatively little atherosclerosis, which led him to conclude that the tortuosity was congenital. The result of a study of two

cases is far from conclusive and furthermore his contention introduces without elucidation the vexed problem: "What is atherosclerosis?" Tortuosity may occur in association with elastic tissue destruction, just as medial and intimal fibrosis may be present in the absence of gross lipid accumulation. In general, tortuosity indicates a loss of elasticity of the vessel wall, and tortuosity could indicate the same thing in the young, particularly in the absence of large-scale, detailed investigations of the histological and physical properties of such arteries. Until results from such a study are forthcoming the hypothesis that even slight carotid tortuosity in the young is a "congenital abnormality" is only for the credulous and another instance of the invocation of the convenient catch-all. One should remember that gross atherosclerosis and hypertension are not essential for the production of tortuosity. Comparison of the tortuosity with that of the splenic artery would be more meaningful.

The frequent coexistence of aneurysms and tortuosity was a striking feature of the angiographic study made by Weibel and Fields.[237] They gave no criteria for aneurysmal dilatation and it is likely that they were in reality viewing evidence of arteriectasis.

Though not universally accepted as such,[237] coiling and kinking are associated with and should be regarded merely as variants of tortuosity, the eventual manifestation probably being dependent on both resilience and function of the perivascular connective tissues.

Absence of the internal carotid artery. Total absence of the internal carotid artery is extremely rare and mostly unilateral.[5,59,107,124,128] Altmann[5] collected 11 instances to 1947, and Lie found 24 cases of the "aplasia." Absence of the artery was bilateral in four instances.

Lie[124] states that the internal carotid artery can safely be assumed to be absent if no bony carotid canal can be found. Arteriography reveals only the presence or absence of a functional lumen, not the presence or absence of the artery. One of the following states[124] may be found:

1. Total absence of one or both internal carotid arteries, or of both internal and external carotid arteries on one side
2. Replacement of the internal carotid by a fibrous cord with or without a small lumen. Histological detail of such cords is not clear. They could be secondary phenomena.
3. Absence of the intracranial segment of the internal carotid
4. Absence of the cervical and petrous segments of the internal carotid
5. Gross narrowing (hypoplasia) affecting the whole internal carotid artery or all of it from a short distance above the bifurcation of the common carotid artery

As a consequence of the absence of an internal carotid artery the corresponding area of supply in the brain is provided with blood from other vessels and secondary compensatory variations occur in these vessels. The area may be supplied from (1) the basilar artery and the contralateral internal carotid artery,[128,225] (2) branches from the internal maxillary, which enter the cranium through the foramen ovale and foramen rotundum to form a common trunk supplying ophthalmic and cerebral branches,[124] (3) the basilar and vertebral arteries entirely via two enlarged posterior communicating arteries, while the ophthalmic arteries arise from the posterior communicating arteries.[59,107]

Lie[124] found incomplete internal carotid arteries on four occasions in the literature. The artery terminated in a large posterior communicating artery in one case[116] and in the other three cases, only the cavernous and cerebral segments were present. The collateral flow was derived from either the contralateral internal carotid, the ophthalmic,[124] or from the multiple anastomotic channels in the petrous portion of the temporal bone together with the ophthalmic artery.[58] In the case reported by Lagarde and colleagues[116] the contralateral internal carotid supplied the area, though their illustration of the topography was unrealistic and the alleged aneurysm of the anterior communicating artery may have been merely a fenestration.

"Hypoplasia" of the internal carotid artery is a term used to describe unusually small internal carotid arteries. Lie[124] reviewed several cases. In a 53-year-old woman, Smith, Nelson, and Dooley[205] reported bilateral "hypoplasia" with a large intracerebral hematoma and small ophthalmic, posterior communicating, middle cerebral, and anterior cerebral arteries. The vertebral, basilar, and posterior cerebral arteries were large, and anastomoses with the meningeal and intracerebral perforating arteries were prominent. The internal carotid arteries abruptly tapered above their origin. Histologically all the great arteries of the neck exhibited intimal fibrosis and atherosclerosis. The history of an unidentified malady at the age of two and the tapering of the arteries are suggestive of an acquired disease of unknown nature.[123] Lie[124] believed the arterial narrowing was caused by involution and that collateral circulation occurs as a compensatory consequence. At this stage it is wisest to be skeptical about aplasia and hypoplasia.

Variation in the size of the cervical segment of the internal carotid artery depends on the area of brain that it ultimately supplies. The presence of a carotid-basilar anastomosis will be reflected in the size of the cervical internal carotid. Such variation was stressed by Lehrer[120] who found differences in the caliber of the vessels on each side to be from 5% to 40% in 25% of 142 patients subjected to bilateral angiography. The caliber of the cervical segment was found to be larger on the side where the anterior cerebral artery was enlarged in association with a small proximal segment of the contralateral anterior cerebral.

Lie[124] recorded a large intercarotid anastomosis and absent cervical and petrous portions of the right internal carotid artery. The large vessel branched from the left carotid siphon, crossed the midline, and continued into the right carotid siphon. It probably represents the enlargement of one of the naturally occurring small intercarotid anastomoses normally found between the two carotid arteries.

Carotid-basilar anastomoses. Carotid-basilar anastomoses are unusual anastomotic channels linking the internal carotid to the basilar artery in the postnatal state in man. They are thought to be universal in horses[51] and caused by the development and perpetuation of vessels that are prominent in the early embryo. They are either absorbed or alternatively persist as small insignificant branches in the adult circulation[154] in which their precise incidence is impossible to determine without careful postmortem angiographic studies in combination with anatomical dissection of the vessels from a large series of cadavers. Gross examples are readily found by angiography despite difficulties inherent in the angiographic recognition of the trigeminal artery.[168] A carotid-basilar anastomosis of small caliber more readily passes unrecognized. It is convenient to divide them into three groups depending on the name of the embryonic vessel from which the anastomosis is derived, that is, primitive trigeminal artery, primitive otic (acoustic) artery, and the primitive hypoglossal artery.

Primitive trigeminal artery. A persistent primitive trigeminal artery is the most common of the three types of carotid-basilar anastomosis.[147,222] Fields, Bruetman, and Weibel[58] found 8 instances while investigating 2000 patients over a 4- to 5-year period.

The primitive trigeminal artery provides blood to the longitudinal neural arteries, which in the 3- to 4-month embryo fuse to form the basilar artery. Its importance is temporary and its function is normally usurped in the 5 to 6 mm. embryo by the posterior communicating arteries from the internal carotid. By the time the embryo is 14 mm. in length, it has disappeared.

The first anatomical report[173] of its persistence was in 1844 and a few isolated cases were published subsequently[23,91,150,215] until Sunderland[217] found three cases in 210 dissecting room cadavers. In 1950, Sutton[219] published the first angiographic description

Table 1-2. Incidence of persistent trigeminal artery

Author	Number of cases	Number of angiograms	Number of dissections	Incidence as percentage
Blackburn (1907)[23]	1	—	220	0.45
Sunderland (1948)[217]	3	—	210	1.43
Harrison & Luttrell (1953)[88]	3	582	—	0.52
Stenvers et al. (1953)[210]	1	750	—	0.13
Gessini & Frugoni (1954)[71]	2	160	—	1.25
Poblete & Asenjo (1955)[171]	2	828	—	0.24
Schaerer (1955)[191]	1	60	—	0.17
Rupprecht & Scherzer (1959)[183]	6	Nearly 3000	—	0.2
Schiefer & Walter (1959)[193]	8	1657	—	0.48
Campbell & Dyken (1961)*[36]	4	1076	—	0.37
Bingham & Hayes (1961)[22]	2	1600	—	0.13
Lamb & Morris (1961)[117]	3	1500	—	0.2
Madonick & Ruskin (1962)[130]	3	3000	—	0.1
Passerini & DeDonato (1962)[164]	9	4000	—	0.23
Gilmartin (1963)[80]	3	2207	—	0.14
Eadie et al. (1964)[52]	17	4500	—	0.38
Fields et al. (1965)[58]	8	2000	—	0.4
Krayenbühl & Yaşargil (1965)[114]	14	7305	—	0.19
Dickmann et al. (1967)[49]	7	2000	—	0.35
Lie (1968)[924]	6	3218	—	0.19
Fields (1968)[57]	9	1600	—	0.6
McCormick (1969)[133]	5	—	2000	0.25

*Campbell and Dyken[36] found trigeminal arteries in 0.1% of angiograms at the Indiana University Medical Center and 3 in 76 angiograms at the New Castle State Hospital.

of the anastomosis. A veritable spate of reports has appeared since. Table 1-2 indicates the frequency of its incidence from the literature. Wollschlaeger and Wollschlaeger[250] in 1964 collected 134 recorded cases. Additional cases to a total of 170 have been reported.[25,50,124,209,245,251]

The primitive trigeminal artery arises from the internal carotid artery at an angle of at least 90 degrees as it enters the cavernous sinus[124] and is proximal to the siphon. It runs posteriorly in the cavernous sinus medial to the trigeminal nerve and either penetrates the sella turcica or perforates the dura mater near the clivus (approximately half of the cases).[110,124] The artery communicates with the basilar artery below the origin of the superior cerebellar and above the anterior inferior cerebellar arteries. Angiographically, the shunt of blood and contrast medium to the basilar circulation in carotid angiography is characteristic,[250] but the shunt varies considerably in size. When large, the trigeminal artery supplies most of the blood to the basilar artery and the inferior half or two thirds of this artery and the vertebrals are compensatorily small. At autopsy the vessel will be seen emerging from the dura and joining the basilar artery. The discrepancy in size between the upper and lower segments of the basilar should alert one to the possibility of finding an anastomosis. If small, the trigeminal contributes little to the vertebrobasilar circulation, and such cases are very likely to be overlooked at both autopsy and angiography.

In one instance a left primitive trigeminal artery supplied its fellow of the opposite side and then both vessels joined the basilar artery.[29] In the remaining reported cases, the artery was unilateral, the right side predominating slightly.[250]

Controversy exists concerning the significance of the primitive trigeminal artery, which is not generally related to any special pathological entity.[124,241,250] Not all instances of this anastomosis find their way into medical literature, and only emphasis on an association with a particular disease entity or a review is likely to be sanctioned for publication. In association with cere-

Table 1-3. Association of persistent trigeminal artery and intracranial arterial aneurysm

Author	Sex	Age in years	Site, trigeminal artery	Site of aneurysm
Slany (1938)[203]	F	48	Right	1. Left middle cerebral 2. Right internal carotid
Poblete & Asenjo (1955)[171]	F	54	Right	Cavernous segment of right internal carotid
Phillipides et al. (1952)[169]	M	53	—	Basilar
Murtagh et al. (1955)[147]	M	62	Left	Left middle cerebral
Schaerer (1955)[191]	M	40	Right	Left middle cerebral
Davis et al. (1956)[47]	—	—	—	Trigeminal
Mount & Taveras (1956)[146]	M	42	Right	Left internal carotid
Wiedenmann & Hipp (1959)[240]	M	40	Right	Right anterior cerebral
Meyer & Busch (1960)[141]	F	24	Right	1. Basilar 2. Right posterior cerebral 3. Anterior cerebral
Brenner (1960)[30]	F	25	Right	Anterior communicating
Lamb & Morris (1961)[117]	F	62	Left	Right posterior communicating
Bingham & Hayes [1961)[22]	M	52	Left	Anterior communicating
Passerini & DeDonato (1962)[164]	1. F*	71	Left	Cavernous sinus
	2. M	27	Left	Anterior communicating
Bossi & Caffaratti (1963)[25]	M	61	Right	Trigeminal
Wise & Palubinskas (1964)[245]	1. F	52	Left	Left internal carotid
	2. F	52	Left	Anterior communicating
Eadie et al. (1964)[52]	(No details of five cases available)			
Djindjian et al. (1965)[50]	1. —	—	—	Anterior communicating
	2. —	—	—	Junction of posterior communicating and internal carotid
	3. —	—	—	Junction of posterior communicating and internal carotid
	4. —	—	—	—
Krayenbühl & Yaşargil (1965)[114]	—	—	Homolateral	Internal carotid and posterior communicating
Wolpert (1966)[251]	1. F	57	Right	Trigeminal
	2. M	35	Left	1. Left posterior communicating 2. Basilar (junction with anastomosis), basilar bifurcation
Lie (1968)[124]	F	64	Right	Anterior communicating
Bull (1969)[32]	F	43	Right	Trigeminal artery (possible)
Stehbens (1970)[209]	M	70	Right	Anterior communicating
McCormick (1970)[134]	M	47	Left	Left internal carotid at origin of ophthalmic
	F	57	Right	Left middle cerebral

*Also reported by Lombardi et al.[127]

bral aneurysms, 36 cases have now been observed (Table 1-3). The presence of the artery is alleged to indicate that mechanical weaknesses, sufficient to predispose the patient to aneurysm formation,[251] may occur in the encephalic arteries. No such deficiency in the wall has been demonstrated. The fact that the primitive artery existed as an endothelial tube in an embryo of some 4 to 5 mm. has been assumed to be sufficient verification of a developmental weakness of the cavernous segment and of a predisposition to the formation of aneurysms and carotid-cavernous fistulas[251] whether they be of spontaneous or traumatic origin. However the age incidence of the onset of cerebral aneurysms with a persistence of the trigeminal artery hardly suggests a developmental disease, and furthermore the few aneurysms geographically related to the artery appear to be of the "arteriosclerotic" type. Indeed the tri-

Table 1-4. Reported instances of persistent trigeminal artery associated with arteriovenous aneurysms

Author	Age in years	Sex	Side of persistent trigeminal artery	Site of vascular lesion
Saltzman (1959)[186]	69	Female	Left	Left frontal (multiple)
Schiefer & Walter (1959)[193]	51	Male	Left	Left parietal
Campbell & Dyken (1961)*[36]	38	Female	Right	Multiple small angiomas related to middle cerebral artery
Lamb & Morris (1961)[117]	23	Female	Left	Left posterior parietal
Gannon (1962)[68]	46	Female	Right	Right parietal
Eadie, Jamieson, & Lennon (1964)[52] Case 1	Information not available			
Eadie, Jamieson, & Lennon (1964)[52] Case 2	Information not available			
Fields (1968)[57]	25	Female (pregnant)	Left	Right parietooccipital

*Patient had history of traumatic birth with right hemiparesis since infancy.

geminal artery is often ectatic and coexistent hypertension not infrequent.

Sutton[219] and others[36,114,124,193] have recorded a persistent trigeminal artery in association with a cerebral tumor. Passerini and DeDonato[164] in 4000 angiograms found five cases associated with cerebral tumors and, although no causative relationship was put forward, their two cases with cerebral aneurysm were emphasized.

Appearing on a so-called negative angiogram, a primitive trigeminal artery has been held to be the cause of subarachnoid hemorrhage.[112,193] But negative angiograms are not infrequent in subarachnoid hemorrhage and to ascribe the hemorrhage to the anastomosis is but speculation and indicative of observer bias. Saltzman[186] reported a case of an arteriovenous aneurysm in the ipsilateral frontal lobe as did Harrison and Luttrell[88] with the added complication of intracerebral hemorrhage. Eight cases of this association are now known (Table 1-4).

Trigeminal neuralgia, presumably caused by an ectatic anastomotic vessel exerting pressure on the sensory branch of the trigeminal nerve,[100,124] has been recorded as has hyperesthesia. In a few instances the ocular nerves have been involved by this artery.[124,130]

Djindjian and colleagues[50] reported thrombosis of the internal carotid artery where a trigeminal artery originated, and this is not surprising in view of the longevity and severity of atherosclerosis in such cases. Fotopulos[66] recorded thrombosis of the contralateral internal carotid, and a few similar vascular occlusions have been observed.[117,124,164] There is no evidence for contending that the shunt results in a relative deficiency in the region of carotid supply,[165] and hemodynamic studies should establish this as fact. Campbell and Dyken[36] felt that differences could exist in retinal and cerebral arterial pressures on the two sides. The results of ophthalmodynometric measurements in two patients were equivocal.

Primitive otic (acoustic) artery. The primitive otic (acoustic or acousticofacial) artery is usually the first of the three primitive presegmental arteries to disappear. It is quite transitory and of little importance in the blood supply to the posterior cranial fossa.[124]

A few cases supposedly denoting the persistence of this artery have been reported,[5,114,240] but Lie,[124] in reviewing the literature, does not accept them and no indubitable case of the persistence of this anastomosis is extant. All attempts to es-

tablish its existence have come to naught. Lie[124] believes that the primitive otic artery should normally arise from the petrous segment of the internal carotid artery and pass with the facial and auditory nerve through the internal auditory meatus turning medially to join the basilar artery between the anterior and posterior inferior cerebellar arteries. If the otic artery is of any size, reduction in dimensions of the vertebral arteries transpires.

Primitive hypoglossal artery. The primitive hypoglossal artery (last occipital or hypoglossal artery[124]), variously held to be single or to consist of 2 or 3 channels, disappears after the otic artery when the embryo is approximately 5 mm long.[124] Regarding the origin of the hypoglossal artery in the adult, Morris and Moffat[145] contend that the primitive lateral basilovertebral anastomoses of Padget are bilateral channels parallel to the longitudinal neural arteries forming, as their name implies, anastomotic vessels between the vertebral and basilar arteries. Morris and Moffat[145] consider that the adult hypoglossal artery is derived from three sources, the first portion being near the carotid artery consists of the primitive hypoglossal artery itself, the second segment is derived from the lateral basilovertebral anastomosis normally communicating with the longitudinal neural arteries (future basilar) by means of transverse anastomosing channels, one of which is thought to constitute the third part. Observations in support of this concept[124] are the following: (1) The postnatal hypoglossal artery is lateral and dorsal to the hypoglossal nerve roots rather than ventromedial as is the embryonic vessel. The anatomical location can be explained only if the lateral basilovertebral anastomosis and a transverse channel to the primitive basilar participate in the eventual development of the hypoglossal artery, (2) The posterior inferior cerebellar artery arises from the lateral anastomotic channel in the embryo and in some instances from the postnatal hypoglossal artery.[145]

Lie[124] found 20 instances of this hypoglossal artery in the adult. Others[58,188] have brought the total to 30; so it is much less common than the trigeminal artery. Gilmartin[80] found two instances in 2207 angiograms, and Wiedenmann and Hipp[240] found two in 7382 angiograms. Most have been demonstrated by angiography,[43,124] and as with the otic artery the authenticity of some reported cases is doubtful.[124] Lie[124] offers four criteria for the diagnosis:

1. The artery should emerge as a robust branch from the cervical segment of the internal carotid artery at the level of the first, second, or third cervical vertebra.
2. After a tortuous course the artery should pass through the anterior condylar foramen (hypoglossal canal) and not the foramen magnum to the posterior cranial fossa.
3. The basilar artery filled just beyond the point of anastomosis with the hypoglossal artery.
4. The posterior communicating arteries should not be visible by angiography.

To compensate, the vertebral arteries are very small. Samra and associates[188] found the hypoglossal arising from the common carotid artery and in Oertel's case[150] it was associated with a persistent trigeminal artery. This anatomical variation is of no great significance[31] and the comment concerning the primitive trigeminal artery is applicable here. Lie[124] considered the variation purely as an accidental finding on arteriograms performed for pathological lesions such as cerebral tumor,[164,196] mental impairment, subarachnoid hemorrhage, and cerebrovascular accidents. Four instances of this carotid-basilar anastomosis in association with a saccular aneurysm have come to light.[118,124,228] Neither histological nor gross examinations of these aneurysms were attempted.

In a mildly hypertensive woman of 57 years dying with multiple cerebellar and occipital lobe infarcts, Gilmartin[80] reported a hypoglossal artery in conjunction with old myocardial infarction. Radiologically there was stenosis of the internal carotid artery with poststenotic dilatation proximal to the origin of the hypoglossal

artery. Lie[124] found associated thrombosis of a middle cerebral artery in a 63-year-old woman. Gerlach and associates[70] reported a traumatic carotid cavernous fistula on the same side as the hypoglossal artery.

Carotid-vertebral anastomosis. The proatlantal intersegmental artery embryologically supplies blood from the dorsal aorta to the longitudinal neural artery and thence to the posterior cranial fossa. The counterpart in the adult is an anastomotic vessel arising from the cervical segment of the internal carotid artery and joining the horizontal segment of the vertebral artery in the neck. Lie[124] found three cases in the literature.

Ophthalmic artery

Anatomy. The ophthalmic artery, the first branch of the cerebral segment of the internal carotid artery, arises under cover of the optic nerve at an obtuse angle from the anteromedial aspect of the artery immediately after the internal carotid emerges from the cavernous sinus and passes through the dura mater. Alternatively it may arise from the cavernous segment passing anterolaterally below the optic nerve and through the optic foramen into the orbital cavity where anastomoses with branches of the external carotid artery are frequent.

Variations. Variations of the intracranial segment are extremely rare but replacement by the branch of the middle meningeal artery has been described[27] as has origin from the middle cerebral in the absence of the internal carotid.[128]

Posterior communicating artery

Anatomy. The posterior communicating artery arises from the dorsal convex surface of the cerebral segment of the internal carotid artery. It proceeds posteriorly and medially below the optic tract and above the oculomotor nerve to anastomose with the posterior cerebral artery at a varying distance from the bifurcation of the basilar artery. It gives off a number of small branches to the optic chiasm and tract, the cerebral peduncle, the internal capsule, the medial surface of the thalamus, and the wall of the third ventricle and assists, to a greater or lesser extent than the posterior cerebral, in the supply of the occipital pole and the tentorial aspect of the cerebrum. When the posterior communicating artery is small, it often appears to diminish in caliber as it proceeds backwards indicating that the flow is in that direction. Angiography supports this observation, for the artery fills more frequently in carotid angiograms than in vertebral.

Kaplan and Ford[105] refer to the posterior communicating artery as the proximal portion of the posterior cerebral and to the small arterial segment that is a terminal branch of the basilar (divisional branch of the basilar) as the mesencephalic artery (Fig. 1-2). Hasebe[90] named the posterior communicating artery the pars carotica and the divisional branch of the basilar the pars basilaris. Such nomenclature unless universally adopted only adds to the general confusion.

Variations. There are frequent variations in the size of the posterior communicating artery and of the divisional branch of the basilar (proximal segment of the posterior cerebral artery). The role played by each of these two arteries in the supply of the distal segment of the posterior cerebral artery seems to be inversely proportional to that of the other and less often the anterior choroidal artery usurps the role of both.

Fetterman and Moran[56] examined the posterior communicating arteries of 200 brains, derived mostly from old individuals. Those circles of Willis in which the posterior communicating arteries contributed most of the blood to the distal segments of the posterior cerebral arteries were referred to as being of the primitive type.[97,189] The circles in which the basilar contributed most of the blood to the distal segment of the posterior cerebral on both sides were said to be normal or average circles. When the posterior communicating