

Microneurosurgery

in 4 Volumes

M. G. Yaşargil

IIIA AVM of the Brain,
History, Embryology,
Pathological Considerations,
Hemodynamics,
Diagnostic Studies,
Microsurgical Anatomy

III A AVM of the Brain, History, Embryology, Pathological Considerations, Hemodynamics, Diagnostic Studies, Microsurgical Anatomy

M. G. Yaşargil

Collaborators: P. J. Teddy, A. Valavanis

Contributors:

H. M. Duvernoy, H. M. Keller,
St. Kubik, M. Mañin-Padilla

Illustrated by P. Roth

242 partly colored Figures in
808 Illustrations, 34 Tables



1987

Georg Thieme Verlag
Stuttgart · New York

Thieme Medical Publishers, Inc.
New York

Addresses

Yaşargil, M. G., M. D., Professor and Chairman
Neurosurgical Department
University Hospital, Zurich

Duvernoy, H. M., M. D., Professor
Laboratoire d'Anatomie
Université de Besançon, Faculté de Médecine
Besançon

Keller, H. M., M. D., Ph. D., Professor of Neurology
Zurich

Kubik, St., M. D., Professor
Institute of Anatomy
University of Zurich

Marin-Padilla, M., M. D., Professor
Department of Pathology
Dartmouth College
Hanover, New Hampshire

Teddy, P. J., DPhil, FRCS, Consultant Neurosurgeon
The Department of Neurological Surgery
Oxfordshire Health Authority
The Radcliffe Infirmary, Oxford

Valavanis, A., M. D., Professor
Department of Neuroradiology
University Hospital, Zurich

Roth, P., Scientific artist
Neurosurgical Department
University Hospital, Zurich

Important Note: Medicine is an ever-changing science. Research and clinical experience are continually broadening our knowledge, in particular our knowledge of proper treatment and drug therapy. Insofar as this book mentions any dosage or application, readers may rest assured that the authors, editors and publishers have made every effort to ensure that such references are strictly in accordance with the state of knowledge at the time of production of the book. Nevertheless, every user is requested to carefully examine the manufacturer's leaflets accompanying each drug to check on his own responsibility whether the dosage schedules recommended therein or the contraindications stated by the manufacturers differ from the statements made in the present book. Such examination is particularly important with drugs that are either rarely used or have been newly released on the market.

Some of the product names, patents and registered designs referred to in this book are in fact registered trademarks or proprietary names even though specific reference to this fact is not always made in the text. Therefore, the appearance of a name without designation as proprietary is not to be construed as a representation by the publisher that it is in the public domain.

This book, including all parts thereof, is legally protected by copyright. Any use, exploitation or commercialization outside the narrow limits set by copyright legislation, without the publisher's consent, is illegal and liable to prosecution. This applies in particular to photostat reproduction, copying, mimeographing or duplication of any kind, translating, preparation of microfilms, and electronic data processing and storage.

© 1987 Georg Thieme Verlag, Rüdigerstraße 14,
D-7000 Stuttgart 30, Germany
Thieme Medical Publishers, Inc.,
381 Park Avenue South, New York, N.Y. 10016
Printed in Germany.

Typesetting: R. Hurler, D-7311 Notzingen,
typeset on Linotron 202;
Printed by K. Grammlich, D-7401 Pliezhausen

ISBN 3-13-645001-9 (Georg Thieme Verlag, Stuttgart)
ISBN 0-86577-258-4 (Thieme Medical Publishers, Inc., New York)

Contents

Introduction	1
1 History	3
A Short History of the Diagnosis and Treatment of Cerebral AVMs	3
Pre-17th Century	3
17th-19th Century	3
Treatment of Extracranial AVM in Earlier and Present Time	5
Intracranial Angiomas	5
The Contributions of Virchow and his Contemporaries	5
Early Clinical Observations on Intracranial AVMs	6
Surgical Treatment of Cerebral AVMs (1889–1930)	7
Neurosurgical Approaches Prior to the Introduction of Angiography (1928)	9
Neurosurgical Treatment of Intracranial AVM Following the Introduction of Angiography (1930)	10
The Limitations of Surgery	12
Conservative versus Surgical Treatment ..	14
Summary and Outlook	20
Diagnosis	20
Surgery	20
2 Embryology	23
<i>Miguel Marin-Padilla</i>	
A. Embryogenesis of the Early Vascularization of the Central Nervous System	23
Introduction	23
Perineural Vascular Territory of the CNS Vasculature	24
Interneural Vascular Territory of the CNS Vasculature	31
Composition and Organization of the Pial Vascular Plexus	31
Vascular Perforation of the CNS Surface by Pial Vessels	32
Vascular Approach and Contact with the CNS Surface	32
Endothelial Filopodia Perforation of CNS Surface	32
In Situ Formation of New Intraneural Vessels	37
Establishment of the VRC and Interneural Vascular Territory	37
Intraneural Vascular Territory of the CNS Vasculature	39
Conclusions	44
B. Vascular Malformation of the Central Nervous System. Embryological Considerations	45
Capillary Telangiectasias and Cavernous Angiomas	46
Venous and Arteriovenous Malformations ..	46
Sturge-Weber-Dimitri's Disease	47

3 Pathological Considerations	49
Pathogenesis	49
Classification of Vascular Malformations	57
Nomenclature	57
Classification	58
The Author's Classification	61
Location of AVMs	63
Localization	63
Localization of the AVM Within the Brain	63
I. Surface Lesions (visible on exploration on the surface of the brain)	64
II. Deep Lesions (invisible at exploration on the surface)	64
The Nidus	73
The Concept of Compartments	74
Compact and Diffuse Lesions	76
Sizes, Shapes, and Elements of AVMs	85
Sizes of AVM	85
Shape	92
Pure Fistulous AVM	100
Elements of an AVM	111
Arterial Feeders	111
Venous Drainage	118
Sinuses	138
Enlargement, Growth, and Regrowth of AVMs	138
Enlargement	140
Growth	146
Pseudo-Growth	146
True Growth of the AVM	155
Spontaneous Thrombosis and Regression of AVMs	161
Multiple AVMs	165
Multiple Cerebral AVMs	165
Intracranial and Intraspinal AVMs	182
Association of Persistent Trigeminal Artery and AVM	182
Association of Aneurysm and AVM	182
Intracranial AVM with Stenosis and Occlusion of Major Vessels	190
Arterial	190
Moya-Moya Disease	192
Venous	193
AVM Associated with Other Pathological Entities	211
Tumors	211
4 Hemodynamics	213
Introduction	213
Hemodynamics	214
The Physics of Fluids and Blood Flow	214
Pressure, Flow, and Resistance	214
The Nature of Blood	215
Laminar versus Turbulent Flow	215
Tortuosity of Vessels	216
Vascular Distensibility	216
Cerebral Circulation:	
Functional Anatomy of the Cerebral Circulation	217
Neuronal Innervation	217
Microcirculation	218
AVM Structure	218
Enlargement of AVMs	218
Autoregulation	220
Normal Perfusion Pressure Breakthrough	221
Comments on the Normal Perfusion Pressure Breakthrough Theory	222
Effects of AVMs upon Cerebral Function	227
Local Mass Effect	227
Obstruction	227
Hemorrhage	227
Vascular Steal	228
Systemic Effects	235
Operative Considerations with Regard to Hemodynamics	237
Preoperative Evaluation	237
Operative Techniques	237
Postoperative Care	239

5 Diagnosis and Follow-up of Patients with Cerebral AVM using Doppler Ultrasound	240
<i>Herbert M. Keller</i>	
6 Neuroradiological Evaluation	250
<i>A. Valavanis</i>	
Computed Tomography	250
Magnetic Resonance Imaging (MRI)	259
Cerebral Angiography	260
Technique	260
Erroneous Findings	260
Angiographic Classification	267
Angiographic Investigation	269
Limitations of Conventional Selective Angiography	276
Venous Phase	280
Associated Aneurysms	280
Spasm	280
Summary	283
7 Microsurgical Anatomy of the Brain	284
Supratentorial Sulci and Fissures	284
Fissures	296
Interhemispheric (Longitudinal) Fissure	296
Sylvian Fissure	298
Transverse Fissure	301
Vascular Patterns Relating to Supratentorial Sulci	304
Infratentorial Sulci and Fissures	312
Organization of the Cerebral Microcirculation	320
The Venous System of the Brain	327
The Veins of Posterior Fossa	332
The Collateral Circulation	333
I Extracranial Arterial Circle	334
II Dural Arterial Circle	334
III Basal Cerebral Arterial Circle of Willis	334
IV Cortical Interhemispherical and Intrahemispherical Arterial Circle	334
V Cerebellar Arterial Circle	334
VI Transcranial Arterial Circle	334
VII Spinal Arterial Circle	336
VIII Vertebro-cervical Arterial Circle	336
IX Primitive Arterial Circle	336
Pathology of Collateral System	337
8 Cortical Blood Vessels of the Human Brain	338
<i>H. M. Duvernoy</i>	
Blood Vessels of the Cerebral Cortex	338
I Pial Vessels	338
The Pial Arterial Network	338
The Pial Venous Network	339
Pial Vessels: Discussion	340
Intracortical Vessels	340
Blood Vessels of the Cerebellar Cortex	345
I Pial Vessels	345
II Intracortical Vessels	347

9 Anatomy of the Calcarine Sulcus	350
<i>S. Kubik and B. Szarvas</i>	
Development	351
Nomenclature of the Various Parts of the Calcarine Sulcus	351
The Location of the Meeting Point	352
Anatomical Variations of the Pars Posterior	352
Variations in the Terminal Part of the Pars Posterior	352
Side-Branches and Connections	355
Pars Anterior of the Calcarine Sulcus	356
Variations in Course	357
Connections and Side-Branches	357
Inner Structure of the Sulcus Calcarinus	357
Measurements	366
The Relationship Between the Sulcus Calcarinus and the Calcar Avis, Posterior Horn and Optic Radiation	366
References	369
Index	401

Introduction

This volume attempts to provide the basis for an informed approach toward operating upon AVMs and for learning the actual operative techniques favoured by the authors. It is not meant to be a comprehensive review of all that has been written in the past upon the subject. While much of the material is based upon the findings and theories of others, some is also new. Historically, the development of new surgical disciplines has usually created a need for ever more detailed study of the embryology, anatomy, physiology and anesthesia relevant to that field. The particular needs of the surgeon have often stimulated new methods of carrying out these studies. Neurosurgery has been no exception. Initially, there was the need for a general understanding of the gross anatomy and the relationships and physiology of fiber tracts, cranial nerves and cortical structures, which would allow the surgeon to operate with relative safety within the limits imposed by the instrumentation, anesthesia, and illumination available at that particular time.

Cerebral angiography has been a real "break-through", not only for diagnostic purposes but also for a better understanding of the hemodynamic and therefore the functional anatomy of the central nervous system. Neuroradiological anatomy, with entirely new perspectives was born and stimulated the neurosurgeon to expand his surgical activities. Refined angiography has permitted accurate study of vessels within the living brain, complementing the work of the pure anatomist. Selective and superselective angiographic techniques have been created as well as interventional neuroradiology. Endovascular neurosurgery was nothing more than the logical consequence of this accelerated development that has occurred within the last 20 years. Again the perspective concerning the anatomy of the central nervous system has been broadened.

The introduction of stereotactic techniques has led to the development of precise atlases of deep areas within the brain (Szikla et al. 1977) and now to computerized three dimensional maps of some

of these structures (Salamon and Huang 1980, Unsöld et al. 1982).

At the same time, microtechniques were introduced into neurosurgery. The ability to reach areas, previously deemed inaccessible, with comparative safety, has dictated the need for a new perspective of the microanatomical and topographical relationship of almost every part of the cranial contents. The work of Basset (1952), Huang (1946–1985), Stephens and Stilwell (1969), Duvernoy (1969–1983), Waddington (1974), Newton and Potts (1974), Williams and Warwick (1975), Lang (1981), Seeger (1978, 1980, 1984), has given us, in large part, the necessary topographical details. The elegant series of studies by Rhoton and his associates (1976–1985) describe precise microsurgical details of various brain areas, with their corresponding vasculature, from the point of view of the neurosurgeon. These neuroanatomical publications offer, besides profound and scientifically proven knowledge, very detailed geometrical, trigonometrical-arithmetical data concerning lengths and diameters of various bony, nervous and vascular structures, as well as distances between them. These painstaking and precise elaborations are essential background information, indispensable for every neurosurgical procedure. These major works, dealing with the brain stem vasculature comprise a precise review of the neurosurgical anatomy of the base of the brain, brain stem and circle of Willis. Our own account of the basal cisterns and circle of Willis has been described in Volume I.

Unlike the great majority of aneurysms, arteriovenous malformations and cerebral tumors are not confined to the basal regions of the brain. A new perspective must therefore be adopted, namely the awareness that even the deepest structures may often be reached by working carefully within the sulci and fissures of the brain. The basic patterns of these important anatomical structures should now be studied. Detailed accounts of sulcal and fissural anatomy are rare and generally incomplete but neurosurgery would certainly benefit from more precise studies in the future.

For this reason it seemed necessary to study, analyze and present the brain anatomy in a new concept, from the view point of the sulcal and fissural systems as well as their relation to the vessels.

Originally we planned to study the detailed anatomy of these systems in collaboration with Professor Kubik of Zurich, and to include these results in the present publication. This undertaking, however, turned out to be much more time-consuming than originally estimated.

The sulcal system showed an amazing degree of variation giving the impression of being a highly irregular system. As this study proceeded, however, it was realized that this irregularity of the sulcal system conforms to certain general principles. Despite the fascinating preliminary observations, we finally decided not to further delay the publication of this volume and to include here only the detailed anatomy of the calcarine sulcus and its variations and to present some representative displays of general sulcal anatomy. From this contribution by Professor Kubik, the reader will certainly become aware of the fascinating world of sulci. This interesting work will be continued and published later.

Although knowledge of sulcal and fissural anatomy is extremely important for angioma and tumor surgery, an equally detailed knowledge of microvascular anatomy is essential in order to perfect microsurgical techniques. Only with this knowledge can the neurosurgeon fulfil his goal, which is to preserve and protect the brain parenchyma adjacent to the lesion.

Since the pioneering work of Heubner (1872) and Duret (1873) on cerebral microvascularization and microcirculation, subsequent generations of anatomists further refined and extended their original concepts. Stimulated by the recent, excellent work of Duvernoy et al. of Besançon, France, we invited Professor Duvernoy to provide a concise chapter on cerebral microvascularization in order to stimulate younger colleagues to pursue the endeavours of modern anatomists.

We have been fortunate to have Professor Marin-Padilla of Hanover/New Hampshire, USA, who has contributed a concise chapter on the embryology of brain vessels, also summarizing the history in this field and adding his new ideas regarding the possible formation of cerebral vascular malformations. The development and maldevelopment of the cerebral venous system was intentionally not included in this volume, since this has been comprehensively described by Huang et al. as recently as 1984.

As we already noted in the first volume on aneurysms, the detailed anatomy of an AVM can

only be completely and definitely evaluated at microsurgical exploration and not by any imaging technique. Although superselective angiography provides essential information regarding the composition of an AVM, we would like to have even more sophisticated angiographic techniques for even more precise study of the vascular composition and the hemodynamics of the AVM nidus and its compartments; this is already practiced daily by interventional neuroradiologists for vascular lesions of the skull base as well as head and neck. Professor Valavanis of Zurich, who performed all pre- and postoperative neuroradiological procedures since 1978, has been invited to provide the chapter on the neuroradiological evaluation of cerebral vascular malformations, also summarizing the relative role of CT, MRI and angiography. We refused to perform invasive studies to assess the hemodynamics of AVMs in our patients. However, we routinely applied non-invasive Doppler-ultrasound pre- and postoperatively. Professor Keller of Zurich, has contributed a separate chapter on his Doppler-ultrasound technique, summarizing the principles and the results of this method.

Modern neurosurgery is inherently dependant on the advances in neuroanesthetic techniques. During the last 20 years, five groups of anesthesiologists were involved in our daily work. The results achieved in the surgical management of intracranial AVMs were also possible thanks to the great effort of our neuroanesthesiologists, especially Drs. M. Curcic and Dr. M. Kis, who have been responsible for neuroanesthesia during the last 10 years.

In Volume III B of this series, the surgical techniques and results as well as the neuroanesthetic technique will be presented in detail.

1 History

A Short History of the Diagnosis and Treatment of Cerebral AVMs

As attested by F. Henschen (1955) angiomatous malformations and tumors have been, since Virchow's time, a "problem child" of pathologists. Hamby (1958) defined the main problems posed in understanding the pathology of these lesions and his statements are valid even today:

"The origin and anatomy of the cerebral angiomas has frustrated pathologists over the years as much as their treatment has baffled surgeons. An extensive literature has developed, replete with picturesque nomenclature based upon attempts to describe the appearance of lesions seen at the operative table or at necropsy. The surgical descriptions are not entirely basic nor accurate because the bulk of the lesion is largely submerged under the cortex and hence invisible to the examiner. The pathologic descriptions have been faulty because of deflation of the lesion at the time of examination by lack of the expansile blood stream that characterizes them in life. Also confusing the picture of the dead lesion are the alterations produced in the component vessels by blood under arterial pressure, which dilates veins and "arterializes" them to withstand the added stress. Vascular resistance being lowered by the shunt, arteries dilate to carry more blood under less than usual pressure, and lose some of their usual characteristics."

However, the introduction of cerebral angiography (Moniz 1927) together with the continuing improvements in the quality of angiograms and the remarkable developments in vascular catheterization techniques (Seldinger 1953, Djindjian 1962) has opened up new dimensions in the study of the morphological and hemodynamic aspects of AVMs.

This short historical review may help to understand how we have arrived at the present day

interpretations of AVM pathology and development and how modes of treatment have evolved.

Pre-17th Century

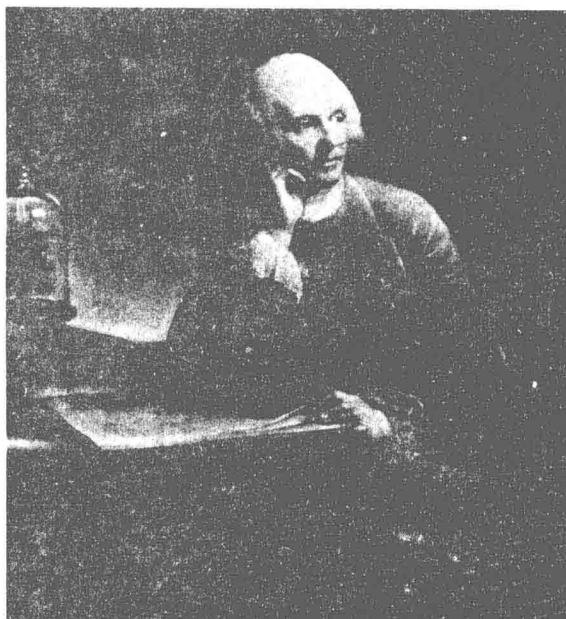
Descriptions of vascular malformations of the skin and other visible organs such as eye, lips and ear with occasional comments about their often ugly appearance and the difficulty or impossibility of treatment may be seen in some of the earliest recorded historical manuscripts.

The Papyrus Ebers (ca. 1500 BC) contained descriptions of hemorrhoids, skin tumors, hydroceles, varicose veins and aneurysms. Kharadly (1956) showed that hernias and aneurysms were operated upon even in those times but not AVMs. The warning, "You must keep your hands off — Noli me tangere" is stated in the relevant chapter.

Virchow cited prominent physicians like Hippocrates, Galen, Celsus, Aetius, Avicenna, and Vidus Vidius, who were dealing with the diagnosis and treatment of different types of external vascular malformations. Von Bramann (1886) showed that Galen and Della Groce knew of varicose pulsating swellings and took them to be simple arterial aneurysms. Osler (1915) noted that references to vascular malformations are to be found in the works of Antyllus (2nd Century) and Abulcasis (10th Century).

17th—19th Century

The great breakthroughs in the understanding of the systemic circulation and of the cerebral circulation were made by Harvey (1628) and Willis (1664) respectively.



William Hunter (1718–1783)
(By kind permission of the President and Council of the Royal College of Surgeons of England)



John Hunter (1728–1793)

The work of Harvey and Willis was subsequently complemented by the discovery of the capillary system by Malpighi (1661) and this paved the way for modern theories regarding the evolution and pathology of AVMs.

In the following century (1757) William Hunter was able to identify the clinical characteristics and some hemodynamic aspects of extracranial AVMs. In "Observations on arteriovenous malformation, London Medical Observations and Enquiries, 1762" he wrote: "Vascular malformations of the extremities are caused by an abnormal communication between arteries and veins." Enthusiastic phlebotomists of that period prepared two perfect examples of arteriovenous aneurysm for W. Hunter, which he was quick to recognize (cit. Dandy 1928); at the point of communication between the artery and the vein, he recognized a loud hissing bruit and a strong tremulous thrill: large tortuous sacs were seen to pulsate; the brachial artery was greatly enlarged and serpentine cephalad to the arteriovenous fistula, but distal to it, the artery became smaller than on the other side. He was able to reduce the size of the vein, stop their pulsation and eliminate both the bruit and the thrill by pressing on a localized spot, which he recognized to be the opening between the artery and vein. It was William Hunter who first suggested the term "anastomosis" to denote the union of the two vessels, whereas the term "collateral" was introduced by his

younger brother John Hunter who also ligated the femoral artery in a case with popliteal aneurysm and proved the efficiency of the collateral arterial system.

The broad scientific approach concerning the nature of these impressive aberrations began with pathologists and surgeons 200 years ago who described them as "erectile tumors" and swellings of the skin and organs. The advent of medical journals enabled the scientists to publish their observations.

After 1850 the number of publications concerning these erectile tumors increased rapidly. Between the time of William Hunter (1762) and Sonntag (1919), 65 such publications are to be found: Plenck (1776), Bell (1796), Cruveilhier (1816), Meckel (1818), Dupuytren (1834), Vidal (1846), Rokitsky (1846), Virchow (1851), Gerdy (1852), Schuh (1853, 1866), Busch (1854), Luschka (1854), Esmarch (1854), Lebert (1857), Bennet (1854), and Alibert (1871). More detailed information regarding these papers may be found in the works of Heine (1869), Weber (1869), Körte (1880) and Heineke (1882). Pathological classification based upon varied anatomical descriptions was already becoming clumsy and confusing. By 1894 Wagner had collected from the current literature 24 different nomenclatures. In parallel with changing pathological concepts, the surgery of extracranial AVMs was undergoing a gradual evolution.

Treatment of Extracranial AVM in Earlier and Present Time

The endeavours of general surgeons in dealing with the dangerous and disfiguring extracranial vascular malformations (scalp, external ear, eyelids, orbits, cheeks, lips, tongue, palate and neck) are most informative for the interested neurosurgeon (Beck, Berger, Billroth, Brodie, von Bruns, Bryant, Busch, Caradec, Clairmont, Dalrymple, Dupuytren, Emanuel, Enderlen, German, Goldmann, Heineke, Krause, Lefort, Lieblein, Nélaton, Pilz, Roth, Russell, Schwalbe, Schwartz). Their methods of treatment have, in the past, included:

- 1) Injection of the lesion with: ferrous chloride, glycerin, tannin, chlorzin, carbonic acid, alcohol,
- 2) Electrocauterization,
- 3) Ligation,
- 4) Extirpation.

The variety of modern therapy of external vascular malformations (Williams 1983) shows that therapeutic difficulties still remain in the treatment of these easily approachable lesions:

- 1) Corticotherapy (new born children),
- 2) Radiotherapy,
- 3) Electrocoagulation,
- 4) Cryotherapy,
- 5) Surgery,
- 6) Use of laser beam,
- 7) Embolization.

As in neurosurgery, the advice of most plastic surgeons is that simple ligation of feeding vessels is inadequate and inadvisable.

Intracranial Angiomas

The Contributions of Virchow and his Contemporaries

After exhaustive research work on cavernomas of the liver, Rokitsky (1842–46) came to the conclusion that these were either benign or malignant tumors independent of the surrounding vascular system. Volume 6 of Virchow's *Archive* (1854) contains 3 remarkable papers: Esmarch (pp. 34–57): "Über cavernöse Blutgeschwülste", Luschka (pp. 458–470): "Cavernöse Blutgeschwülste des Gehirns" and Virchow (pp. 526–554): "Über cavernöse (erectile) Geschwülste und Teleangiectasien." Esmarch und Luschka fully supported the neoplasia hypothesis of Rokitsky.

Luschka provided one of the earliest descriptions of an intracranial arteriovenous anomaly in a patient with a frontal cavernoma.

Luschka recognized two types of "Blut-Geschwülste":

- 1) Telangiectases (non neoplastic) arising due to a metamorphosis of capillary systems.
- 2) Cavernous tumors (neoplastic) containing large blood-filled compartments.

The young Virchow, who was involved with research into infection of blood vessels, also published (1851) a remarkable paper concerning "the dilatation of small vessels". In this paper he described and discussed thoroughly his own observations and thoughts and clearly refuted the hypothesis of Rokitsky. In 1863 Virchow published a comprehensive study which may be called the first real milestone in the history of the AVM. In the 3rd volume of his monograph, 200 pages (pp. 306–496) are devoted to the phenomenon of the physiological and pathological changes of blood vessels in all organs. His descriptions profoundly contradicted contemporary opinion. He described telangiectases, venous, arterial, arteriovenous and cystic angiomas (nowadays angioblastomas), and their transitional types, and discussed in detail the pathogenesis of these malformations. He reflected on the atlas of Cruveilhier and the pioneering work of John Bell (*The Principle of Surgery*, London 1826, Volume 3, pp. 326–383, First Edition, London 1796). Bell described cavernoma, AVM and angioblastoma but gave all of them the nomenclature of "Aneurysm per Anastomosis". Virchow said: "This description is still perfectly valid. (p. 328): Aneurysm per anastomosis is an entire change of structure; it is a dilatation of veins, in which they are forced and enlarged by the diseased action of their corresponding arteries. Those happen in consequence of original malformation, a violent action of arteries, and a mutual enlargement of arteries and veins, while the intermediate substance of the part is slowly distended into large intermediate cells, which are dilated to formidable reservoirs of blood. — The blood is poured into the cells of such a tumor by innumerable arteries: from these the blood is continually following into veins, which receive it with such patent orifices etc. The veins form a conspicuous part of such a tumor, but the intermediate cells are an appreciable part of the structure ... (p. 397). All this proves that it is a tissue of small arteries and veins; it fills not like a varix slowly; its filling is by distinct thrombs; it is filled by its small and numerous arteries, and its swelling is (like the erection of the penis) produced by the pulsation of

the arteries, stroke after stroke, pouring out their blood into cells."

"The tumor is a congeries of active vessels and the cellular substance through which these vessels are expanded, resembles the cellular parts of the penis, the gills of a turkey cock or the substance of the placenta, spleen worms."

It is interesting to speculate as to whether Bell was describing a cavernoma or an AVM. It certainly sounds like the modern description of an AVM and its nidus.

Virchow (1854), cited Gerdy (1852), who differentiated eight types of "erectile tumors" and noted the great number of publications concerning the "erectile tumors" and the difficulties with their classification. He preferred the term of "angioma" which was introduced by J. Hughes Bennet (1854) instead of the term of "angionoma" which was advocated by Follin (1861). He credited to Plenck (1776) the term "cavernoma", a nomenclature well recognized in the German literature (Meckel 1818).

Cushing and Bailey (1928) concluded, quite wrongly, that Virchow believed in the neoplastic nature of vascular malformations as proposed by Rokitsky. This error was most likely due to difficulties with translation of the original papers.

Virchow (1863) divided angiomas into cavernous, simple telangiectatic, racemose, and lymphatic types. Racemose angiomas were divided further into arterial and venous types.

Page 474 of Virchow's 3rd Volume (1863) relates to a case of a large extracranial parietooccipital AVM in a man from Florence and described by Vidus Vidius in 1665. Virchow commented that this type of malformation originates through accommodation between artery and vein with consequent dilatation of them (arteriectasie and phlebectasie, p. 471). They are of congenital origin (p. 475). They may grow or spontaneously regress (p. 482). The following nomenclature has been used: aneurysma per anastomosis (Bell) or aneurysma anastomoseon (v. Walther), aneurysma per transfusionem (Dupuytren 1834) and other authors used the term of aneurysma arteriovenosum, or aneurysma varicosum.

Virchow argued (p. 472) that aneurysms would not arise as a result of arteriovenous communication in traumatic cases, therefore the best term would be *aneurysma spurium arterio-venosum*.

Virchow's main concern was not so much nomenclature as the pathophysiology of the lesions. The founder of cellular pathology had a profound interest in pathophysiology. He performed injection studies on the pregnant uterus and placenta and was fascinated by the temporary but enor-

mous increase in capacity of vessels during gestation. In 1851 he spoke of "The physiologic paradigm in the corpora cavernosa of sex organs and the paradigm of pathology in cavernoma and telangiectasis", and further questioned as to whether one type of angioma can transform into another by changes in flow and pressure or by cellular proliferation.

Early Clinical Observations on Intracranial AVMs

Pfannenstiel (1887) and Kaufmann (1897) observed young (22 and 23 years) primipara patients, who died with acute cerebral symptoms. Autopsy study showed a ruptured varicose anomaly of left thalamus opticus and the vena Galeni in one case, and a ruptured varicose anterior callosal anomaly in another.

D'Arcy Power (1888) found, a large AVM in the left sylvian fissure at autopsy on a 20-year-old man who had suffered a hemiplegic stroke and died.

Steinheil (1894) described the history and pathologicoanatomical findings in a patient (59 years) with a large right frontal AVM which drained partially to the vein of Galen. He may thus be credited as being among the earliest to describe the symptomatology of the disease.

Rizzoli 1873 observed a right occipital pulsating swelling in a 9-year-old girl. The pulsation disappeared on compression of the left occipital artery. The girl died from an apparent meningitis (perhaps, in fact, from an intracranial hemorrhage). At autopsy she was found to have an AVM of the occipital region (-duralpial) with drainage to the transverse sinus. There was a defect in the occipital region of the skull so that the pulsation in the AVM could be felt externally.

The first clinical diagnosis of a cerebral AVM was made by Hoffmann (1898). Isenschmid followed the history of this patient, who was presented to medical colleagues in Heidelberg, and discussed the differential diagnoses (1912). He pointed out that the clinical diagnosis of cerebral angiomas had never before been made.

With the onset of operations for brain tumors around 1890, the number of cases of AVM observed clinically, pathologically and surgically began to rise sharply. At that time, contralateral parietal craniotomy for cases of Jacksonian epilepsy occasionally produced an unexpected AVM. Between 1890 and 1936 there were more than 90 reports of around 120 cases of cerebral AVMs. In the cases of Rizzoli (1873), Hoffmann (1898), Isenschmid (1912), Haenel (1926), Eimer and Mehlhose (1927) and in some of the cases of

Dandy (1928) and Cushing and Bailey (1928) the diagnosis was made clinically.

The list of authors who published cases of AVM prior to the angiographic era includes:

Morris (1871), Rizzoli² (1873), von Braman (1886), Pfannenstiel (1887), D'Arcy Power (1888), Giordano¹ (1890), Guldenarm and Winkler¹ (1891), Péan¹ (1891), Starr and McCosh¹ (1894), Steinheil (1895), Lucas-Championnière¹ (1896), Kaufmann (1897), Emanuel (1898), Hoffmann² (1898), Ribbert (1898), Beadles (1899), Shoyer (1900), Struppler (1900), von Bergmann¹ (1901), Chipault¹ (1902), Deetz (1902), Rotgans and Winkler¹ (1902), Kreutz (1903), Bail (1904), Drysdale (1904), Heitmüller (1904), Simmonds (1905), Strominger (1905), Sternberg (1905, 1907), Falk (1906), Lavillette¹ (1906), Dürck (1907), Enders (1908), Krause¹ (1908), Stertzing (1908), Leischner¹ (1909), Ranzel (1909), Tuffier¹ (1909), Blank (1910), Therman (1910–13), Znojensky¹ (1910), Abrikosoff (1911), Astwazaturoff (1911), Cassirer and Mühsam¹ (1911), Isenschmid² (1912), Schmolck (1912), Wichern (1912), von Eiselsberg and Ranzi¹ (1913), Kaiserling (1913), Wischniewski (1913), Castex and Bolo (1914), Leunenschloss (1914), Maklakow¹ (1914), Orbison¹ (1915), Versé (1918), Bort (1920), Castex and Romano (1920), Schmidt (1920), Bannister (1921), Hammes (1921), Magnus¹ (1921), Nonne¹ (1921), Campbell and Ballance¹ (1922), Deist (1922), Worster-Drought and Ballance (1922), Müller (1923), Wohak (1923), Elkin (1924), von Lehoczy (1924), Mühsam¹ (1924), Rienhoff (1924), Esser (1925), Federoff and Bogorad (1925), Klimesch (1925), Laves¹ (1925), Marx (1925), Reid (1925), Dowling (1926), Globus and Strauss (1926), Haenel² (1926), Klimesch (1926), Leeser (1926), Bregman (1927), Eimer and Melhose (1927), Herzog (1927), Olivecrona and Lysholm¹ (1927), Perthes¹ (1927), Worster-Drought and Dickson (1927), Buckley (1928), Cushing and Bailey^{1,2} (1928), Dandy¹ (1928), Ruehl (1929), Yates Paine Brockman^{1,2} (1930), Brock and Dyke (1932), Krug and Samuels (1932), Dimitri and Balado (1933), Levine (1933), Love (1933), Schaltenbrand (1938), Sattler (1939).

¹ Diagnosis made at exploration

² Diagnosis made clinically rest:

Diagnosis made at autopsy

Surgical Treatment of Cerebral AVMs (1889–1930)

Most of these early procedures were carried out by general surgeons (Table 1.1). Giordano is credited to have operated upon the first cerebral AVM in 1889. Regarding his original paper, however, it is clear that he simply ligated a pathological vessel on the left parietal surface and did not expose the remainder of the AVM located in the deep subcortical tissue.



Jules Emile Péan (1830–1898)

(By kind permission of Prof. H. M. Koelbing, Director of the Institute of Medical History, University of Zurich)

The first complete excision of a cerebral AVM was made 98 years ago by the famous French surgeon Péan. He treated a 15-year-old boy who had suffered a left sided Jacksonian fit, and made a diagnosis of a right sided central tumor. The operation took place in May 1889 and was described thus by Péan:

“Au cours de l'opération, nous nous trouvâmes en présence d'un angiome des méninges en communication avec les sinus longitudinal supérieurs. Malgré sa richesse vasculaire, malgré son étendue, la tumeur put être enlevée en totalité, sans perte de sang, grâce au pincement temporaire et définitif des vaisseaux variqueux, dilatés, érectiles, dont elle était composée. A ce propos, nous avons recherché, dans la science les faits de ce genre, qui avaient été publiés et nous n'en avons trouvé

aucun qui fut exactement semblable, aucun surtout qui eût été opéré." Péan's conclusion is optimistic: "— De même qu'il existe des angiomes extracrâniens communiquants à travers la voute du crâne avec le sinus longitudinal supérieur, il existe une variété d'angiomes intracrâniens communicants également avec les sinus longitudinal

supérieurs, mais développés dans l'épaisseur des méninges et situés entièrement à l'intérieur du crâne. Les tumeurs sont justiciables de la trépanation, l'hémorragie et notamment celle due à la communication avec les sinus, et facilement arrêtée par le pincement temporaire et définitif."

Table 1.1 General surgical operations for cerebral AVM

		Localization	Operation	Follow-up
Péan	1889	R central	Extirpation	good
Giordano	1889	L parietal	Ligature of vein	good
Guldenarm	1890	R parietal	Partial extirpation	no follow-up
Guldenarm	1891	R parietal dura-varix	Ligature Extirpation	good
Starr and McCosh	1894	L parietal small angioma	Extirpation	good
Lucas-Championnière	1896	R parietal	Extirpation	good
Rotgans et al.	1897	R parietal	Partial extirpation	good
Rotgans et al.	1898	L parietal	Ligature	good
Chipault	1897–1898	L parietal	3 operations, partial	no follow-up
Von Bergmann	1901	L frontoparietal	Ligature	death, hemorrhage
Lavillette	1906	R parietal	Ligature	no follow-up
Krause	1907	L parietal	Ligature	good
Von Eiselsberg and Ranzi	1907–1908	R parietal (2 cases)	Ligature	good
Leischner	1907	L central	Ligature	good
Tuffier	1909	L central	Ligature	good
Znojensky	1910	Cerebellum	Exploration	death, hemorrhage
Cassirer and Mühsam	1910	R frontoparietal	Extirpation	good
Von Eiselsberg and Ranzi	1913	R paracentral	Ligature of vein	unchanged
Wischnewski	1913	R parietal	Ligature	good
Magnus	1914	L central	Exploration	x-ray, good
Maklakow and Minz	1914	Cerebellum	Exploration	death
Orbison et al.	1915	—	—	—
Nonne	1921	L parietal	Partial extirpation	good
Campbell and Ballance	1922	R parietal	Ligature 2 operations	hemiplegia
Laves	1922	L sylvian fissure	Ligature	death, hemorrhage
Perthes	1923	R parietal	Ligature	good
Yates and Paine	1930	V. Galeni	Exploration	death
Brock and Dyke	1932	R frontoparietal	Exploration	x-ray, good

The original descriptions of these impressive and dramatic attempts to remove cerebral AVMs make fascinating reading.

Neurosurgical Approaches Prior to the Introduction of Angiography (1928)

Cushing (1909–1928) and Dandy (1921–1926) each described their operative experiences in 14 and 15 cases respectively, of venous and arteriovenous malformation and added cases from the literature. Both of their series were published in the same year (1928) and reading the original descriptions it seems likely that all their cases were true arteriovenous malformations. Dandy felt that the only way to cure an arteriovenous aneurysm was to ligate the entering arteries or to excise the whole vascular tumor. Earlier, he had lost one patient from hemorrhage during the operation and a second case from intracerebral hemorrhage following total extirpation and he wrote: "But the radical attempt at cure is attended by such supreme difficulties and is so exceedingly dangerous as to be contraindicated except in certain selected cases... As in most cerebral lesions, however, each case should be considered a law unto itself. There are large aneurysms and small ones; those which are mostly arterial, others mainly venous; some are superficial, others deep, some are in highly important areas of the brain, others in portions largely silent. All of these factors, and finally the patient's wishes in the matter, must be weighed. An aneurysm in the left cerebral hemisphere in a right handed person is surely *noli me tangere* under all conditions. Any attempted cure, even if successful, would almost surely result in disturbances of speech or motor power, or of both... there is more reason to attempt to cure a patient who has an arteriovenous aneurysm in the right hemisphere."

Cushing's experience with operations for cerebral angiomas dated back to 1909. Some brief extracts from his excellent operative accounts follow:

Case 1: A 39-year-old patient presented with raised intracranial pressure thought to be due to a cerebral neoplasm and was operated upon on 3.2.1909: "Left subtemporal decompression was made... The dura was not particularly tense. When opened a large thin-walled venous lake was disclosed, from which branches spread in various directions... It seemed unwise to attempt it."

Case 2: A 4-year-old child with right sided congenital exophthalmos and bulging in the right temporal area; September 4, 1920: "When the dura was reflected there came into view a mass of hugely dilated vessels, evidently veins, which covered the entire temporal lobe. Two of the main vessels were ligated but extirpation was obviously impossible."

Case 3: 30-year-old male, operated on March 18, 1921: "A left osteoplastic exploration was made. When the dura was opened an enormous tangle of dilated veins was disclosed spreading upward from about the region of the arm-center. The larger vessels were fully as big as the little finger. The chief emerging vein was ligated but all attempts to get beneath or between the larger vessels were accompanied by so much bleeding that their ligation or extirpation was deemed impossible."

Case 4: "April 25, 1921: ... On reflecting the dura an exceedingly wet brain was disclosed with two huge veins on the surface, one running largely in the sylvian fissure. The other, more vertical, lay in the precentral fissure... Since the operator felt some regrets at not having been more radical in his attacks upon the lesion in the preceding case, a ligature was first thrown around the large descending vein at the point... A second ligature was then put on, which must have started trouble from stasis in the main varix which became hardened and swollen... Finally bleeding began to occur from around the sides of the varix and a rupture seemed imminent. There was evidently only one thing to do – to catch the base of the protruding lesion with a large curved clip and to throw a ligature around the whole mass. This desperate step was taken and the cavity, which continued to bleed after the ligature was placed, was finally filled with a slab of muscle taken from the patient's leg, before the excessive venous hemorrhage could be controlled. There had been a sharp fall in blood pressure from which she finally recovered without transfusion... As was to be expected, the patient showed a postoperative right hemiplegia and aphasia... Nearly seven years since her operation, regards herself, aside from some remaining weakness of her right arm, as in normal health."

Depressed following such an experience Cushing wrote:

"One could hardly have chosen a worse place than over the lower motor area of the leading hemisphere in which to attempt the surgical removal of a racemose varix." The untoward results of the procedure in this case resulted in a more cautious attitude when a similar lesion was disclosed in the next patient (left postcentral region): "December 28, 1922: No attempt was made to treat the lesion by ligature or otherwise."

Cushing reviewed the poor results of other workers and warned: "The surgical history of most of the reported cases shows not only the futility of an operative attack upon one of these angiomas, but the extreme risk of serious cortical damage which

is entailed... How many less successful attempts, made by surgeons less familiar with intracranial procedures, have gone unrecorded may be left to the imagination."

"The lesions, in short, when accidentally exposed by the surgeon, had better be left alone, and how much radiation may accomplish for them is undetermined though there are favourable experiences on record. So long ago as 1914 Wilhelm Magnus of Oslo unexpectedly exposed at operation a venous angioma of the left rolandic region, a decompression was made with the intention of treating the lesion with radium therapy which at that time was known favourably to influence cutaneous angiomas. After treatment, the decompression, which was bulging, receded, and the epileptiform attacks, from which the patient was suffering, became infrequent and finally disappeared..."

The publication of Reichert (1946) is unique, as he reported 15 cases of premotor vascular anomalies causing Jacksonian epilepsy, which were treated successfully by coagulation of the dural and pial vessels of the lesion (1935 to 1941).

Neurosurgical Treatment of Intracranial AVM Following the Introduction of Angiography (1930)

As we have seen, surgical excision of AVMs was carried out between 1889 and 1930, both by general surgeons and neurosurgeons. Some of these cases met with success, others ended disastrously. After one or two bad results most surgeons did not risk further attempts at excision.

With the advent of cerebral angiography the position began to change, for it became possible not only to diagnose the AVM but also to obtain some idea as to its location, its size and construction and the number of feeding and draining vessels. Angiography, however, was still somewhat primitive and the contrast material imperfect. Only a few angiographic demonstrations of cerebral AVMs were published before 1936 (Dott 1929, Löhr and Jacobi 1933, Moniz 1934 and 1951, Olivecrona and Tönnis 1936). Dott provided the first demonstration of the angiographic aspects of cerebral AVMs at the Neurosurgical Conference in Stockholm in 1935. However, the full benefits of cerebral angiography came only with improved techniques which were not widely available until the 1950s.

Olivecrona had a disappointing experience in 1923 when exploring for an infratentorial tumor (case 65). He was confronted with a highly vascular AVM and the patient died. In another case (66),

Left carotid angiogram showing a frontoparietal AVM. In the monograph of Egas Moniz, "L'Angiographie Cérébrale" Masson, Paris 1934.

