Clinical syndromes of arteriovenous malformations of the transverse-sigmoid sinus¹

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SYNOPSIS Arteriovenous malformations or fistulae shunting arterial blood from branches of the external and internal carotid and vertebral arteries into the transverse-sigmoid sinus may produce different clinical syndromes. The literature is reviewed with 96 patients including six personal cases. Usually these malformations have a congenital origin and only in 4% of the series was there a previous history of a severe head injury. Clinical groups are defined and the role of angiography assessed. Direct surgical approach with occlusion or removal of the vascular malformation is the treatment of choice. Possible methods of treatment by selective embolization are discussed.

During the past 20 years an interesting condition named intradural arteriovenous malformation (AVM), arteriovenous fistula (AVF), or arteriovenous aneurysm (AVA) of the region of the transverse-sigmoid sinus has been recognized.

According to some reviews (Laine *et al.*, 1963; Verbiest, 1968a, b; Aminoff, 1973) there were a few early observations in the 1930s of such dural vascular malformations sometimes also involving the petrous bone and extracranial soft tissue. The two cases of Tönnis (1936) from Olivecrona's series and the patient of Röttgen (1937) demonstrated by angiography are examples of these intradural aneurysms in adults.

Verbiest (1951) published a case that developed an intracranial bruit after a head injury and at operation found an AVA of the tentorium and sigmoid sinus but the angiography did not show the vascular malformation. The same year Obrador and Urquiza (1951) reported the first case of a large AVM of the transverse-sigmoid sinus demonstrated at angiography. It was accompanied by a severe bruit and produced an infantile hydrocephalus.

Since then other cases have been recorded in the literature under different names: intradural

AVA or angiomas; dural AVM of the tentorium, posterior fossa or sigmoid sinus; tentorial AVM, AVF or communications between middle meningeal artery, occipital artery, or external carotid artery with the transverse sinus.

As has been properly remarked by Houser *et al.* (1972), these lesions were rarely identified before 1960 and are emerging now as distinct entities due to the great advances of angiography. These authors also estimate that the various types of dural AVA represent about 10 to 15% of all the intracranial AVA, though they found only 65 cases reported in the literature at the time of their review (Houser *et al.*, 1972).

A careful revision of the literature has been carried out and 96 cases collected, including six personal observations. (see Appendix Table). Four of these have been reported earlier especially from the angiographic point of view (Fernández Urdanibia *et al.*, 1974).

In most of the reported cases of dural or intradural AVM or AVF there is not enough information for an adequate grouping in separate clinical entities. Thus, only some of the collected observations can be considered in the evaluation of certain features.

These AVM or AVF have been related by Aminoff (1973) to the main drainage sinuses. The cases with an anterior and inferior drainage

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TABLE 1
CLINICAL FINDINGS

Case no.	Sex	Age of onset (yr)	Length of history	Symptoms		Neurological examination	Skull radiograph	EEG
				First	Other			
1	M	57	3 m	Headaches, mental deterioration	Fits, hypersomnia, gait unstable	Stuporous, papilloedema, L hemiparesis, bilateral tremor	Normal	R frontotemporal focus, centrencephalic dysfunction
2	M	43	Hours	Headaches, L hemiparesis	—	Disorientation, papilloedema, L hemiparesis, hemihypaesthesia	Increased vascular channels	R frontotemporal focus
3	F	48	12 yr (after head trauma)	Headaches	Bruit, loss of vision, tinnitus, gait unstable	Bruit, papilloedema, nystagmus	Increased vascular channels	L temporal focus
4	F	60	15 d	Headaches, vomiting, loss of vision, bruit (SAH)	—	Bruit, meningeal irritation, L homonymous hemianopsia, hemiparesis, hemihypaesthesia, nystagmus	Increased vascular channels	L temporal focus
5	F	59	8 m	Loss of memory, disorientation	Fits	Mental deterioration, papilloedema, R hemiparesis	Normal	Diffuse cortical and centrencephalic dysfunction
6	F	54	9 m	Tinnitus	Headaches, diplopia, loss of vision	Papilloedema, VI nerve paresis	Normal	Normal

TABLE 2
ANGIOGRAPHIC FINDINGS

Case no. and site	Arterial supply	Primary venous drainage	Anterograde venous flow (along usual channels)	Retrograde venous drainage	Sinus obstruction
1 R TS	Meningeal branches of R occipital artery. Posterior meningeal branch of R vertebral artery	R TS	—	Cortical veins. Straight sinus-internal cerebral veins-basal veins	R SS
2 R TS	Meningeal branches of R and L occipital arteries, R marginal tentorial artery	R TS	—	Cortical veins. Straight sinus-internal cerebral veins-basal veins	R SS
3 R and L TSS	Meningeal branches of R and L occipital arteries. Posterior branches of R and L middle meningeal arteries. Lateral tentorial arteries of R and L internal carotid arteries. Deep cervical artery of the R subclavian artery	R TSS L TS	L jugular vein	—	—
4 R TS	Meningeal branches of R and L occipital arteries. Posterior branches of R middle meningeal artery	Cortical veins R TS	R jugular vein	—	—
5 R TSS	Meningeal branches of R and L occipital arteries. Posterior branch of L middle meningeal artery	L TSS	—	L vein of Labbé-superficial middle cerebral vein-cavernous sinus	Distal portion of SS
6 R TSS	Meningeal branches of R occipital artery. Lateral tentorial artery of R internal carotid artery	R TSS	—	—	Distal portion of R SS

T: transverse. S: sinus. SS: sigmoid sinus.

receive the blood from both carotid arteries into the cavernous, intercavernous, sphenoparietal, and superior or inferior petrosal sinuses. There are several observations of such arteriovenous connections but much more frequent are the AVF of the superior-posterior group of Aminoff

(1973) draining into the superior and inferior sagittal sinuses or the straight, transverse, sigmoid, and occipital sinuses.

Of these groups of dural vascular malformations, the more frequent and of greater clinical interest are the AVF or AVM that shunt and

drain the blood from different branches of the external and internal carotid arteries and vertebral arteries into the transverse-sigmoid sinus. The haemodynamic changes induced by these lesions at the region of the most important out-flow of intradural blood may give rise to a great variety of symptoms and signs.

PERSONAL MATERIAL

Tables 1, 2, and 3 summarize the clinical and angiographic findings as well as the spontaneous evolution or the course after different therapeutic measures in the six cases observed in recent years.

TABLE 3
RESULTS OF SURGICAL TREATMENT

Case no.	Surgical treatment	Follow-up
1	Ligation of branches of external carotid arteries	Died 9 d later
2	—	Hemiparesis
3	Bilateral external carotid artery ligation	Headaches disappeared, bruit persists
4	Occipital craniotomy. Ligation of arterial supply. Drainage of intracerebral haematoma	Complete recovery
5	Ventriculo-peritoneal shunt	Died 13 d later
6	—	Headaches improved, papilloedema continues

It is interesting that five of these patients (cases 1, 2, 3, 5, and 6) presented a clinical syndrome with increased intracranial pressure and papilloedema and that only one of them (case 3) had an audible bruit. Case 4 presented a sub-arachnoid haemorrhage with a bruit and signs of an intracranial vascular lesion.

AETIOLOGY AND PATHOLOGY

Most authors consider these dural AV fistulae as congenital malformations between the arterial branches and the basal emissary veins that drain the extracranial structures into the dural sinuses (Takekawa and Holman, 1965; Newton and Greitz, 1966). The presence of these lesions in small children also favours their congenital

TABLE 4
MAIN SYMPTOMS (92 CASES RECORDED)

Symptom	Total		First symptom	
	(No.)	(%)	(No.)	(%)
Cranial bruit	62	67	43	47
Headaches	46	50	17	18
Subarachnoid haemorrhages	19	20	(4 with bruit)	13
Seizures	14	15	4	4
Tinnitus	14	15	10	11
Visual failure	12	13	2	2
Mental deterioration	11	12	4	4
Exophthalmos, proptosis, ocular congestion and pain	9	10	4	4
Hemiparesis	5	5	3	3
Gait disturbance	4	4	—	—
Vomiting	5	5	1	1
Diplopia	3	3	1	1
Speech disturbance	3	3	1	1
Transient ischaemic episodes	3	3	3	3
Vertigo	2	2	—	—
Hydrocephalus	2	2	(with bruit)	2
Hypacusis	1	1	—	—
Heart failure	1	1	1	1
Dilated scalp veins	1	1	1	1

TABLE 5
CLINICAL SIGNS (91 CASES RECORDED)

(No.) (%)		(No.) (%)	
Bruit	62 68	Increased scalp veins	3 3
Papilloedema	22 24	VII nerve palsy	3 3
Meningeal signs	12 13	Palpable pulsatile mass	3 3
Hemiparesis	11 12	VI nerve palsy	2 2
Hemianopsia	5 5	Nystagmus	2 2
Diplopia	5 5	Ataxia	3 3
Hydrocephalus	4 4	Tremor	1 1
Exophthalmos	4 4	Heart failure	1
Proptosis	4 4	III nerve palsy	1
Amblyopia	5 5	Loss of hearing	1
Cerebellar signs	4 4	V nerve palsy	1
Impairment of consciousness	4 4	Optic atrophy	1
Mental disorders	4 4	Brain-stem syndrome	1

origin as in the cases of Obrador and Urquiza (1951), van der Werf (1964), and others.

Arteriovenous malformations of the intracranial dura mater may also appear in routine necropsies of patients without any neurological complaints. McCormick and Boulter (1966) described two examples of this kind with angiomatous masses in the region of the tentorium and torcular Herophili and mentioned another three cases of such dural angiomas recorded in the literature. These AVM may also be an inci-

TABLE 6
GROUPING OF CLINICAL SYNDROMES
(76 CASES RECORDED)

Group	Signs and symptoms	(No.)	(%)
I	Only subjective complaints due to cranial bruit and tinnitus secondary to AV shunt (A)	27	35
II	Neurological signs due to cerebral ischaemia (B) secondary to AV shunt (A)	8	10
III	Headaches, papilloedema, and visual failure due to AV shunt (A) and increased venous pressure (C)	13	17
IV	Infantile hydrocephalus with bruit due to factors A and C	3	4
V	Subarachnoid haemorrhages due to ruptured pial vessels	13	17
VI	Combination of previous groups	II and III	10
		II and V	1
		III and V	1

dental and asymptomatic finding during angiographies carried out for other cerebral conditions (Aminoff, 1973; Aminoff and Kendall, 1973).

According to Houser *et al.* (1972) nearly all these fistulae are direct communications from dural arteries to dural veins or to both dural and pial veins. Aminoff (1973) also emphasizes this direct communication of the arterial branches of the external and internal carotid arteries with dural veins and sinuses. Their higher frequency along the cranial base and tentorial region may perhaps be explained by the delay in the embryological development of the external carotid artery and by the existence at this location of a great number of emissary veins (Takekawa and Holman, 1965; Houser *et al.*, 1972).

Angiography also visualizes the direct communication between the arteries and venous sinuses without an interposed capillary system (Debrun and Chartres, 1972). In some cases some interposed and dilated arteriovenous sacs or channels joined the dural arteries to the pial veins (Houser *et al.*, 1972). Histological study of these dural AVM reveals masses of multiple and tortuous venous channels of various sizes (Debrun and Chartres, 1972; Aminoff, 1973).

Although these AVM usually are entirely dural they may also have a cerebral pial component (Verbiest, 1968a, b; our case 4) and, as has been properly emphasized by Debrun and Chartres (1972), unless a direct surgical verifica-

TABLE 7
NINETY-SIX ARTERIOVENOUS MALFORMATIONS
OF TRANSVERSE-SIGMOID SINUS

Sex and duration of symptoms	
Sex (recorded in 95 cases)	(No.)
Male	47
Female	48
Duration of symptoms (yr) (recorded in 66 cases)	(Cases)
< 1	43
1-2	5
2-3	6
3-4	2
4-5	2
5-6	1
7-8	1
8-9	1
> 10	5

tion has been performed it is difficult to state whether involvement of the cerebral vessels through corticomeningeal anastomoses is present. In the study of two surgical specimens Houser *et al.* (1972) demonstrated that the AVM drained by pial veins formed a dilated vascular sac lying in the leptomeninges and associated with intracranial haemorrhages. In one of these two cases the haemorrhage extended not only into the subarachnoid and subdural spaces but also into the cerebral parenchyma.

FIG. 1. Case 5. (a) *Left common carotid angiogram. Opacification of a short segment of the left sinus (large arrow) from branches of the left occipital artery and the posterior branch of the left middle meningeal artery (long arrows).* (b) *Retrograde flow into the left vein of Labbé (arrow) from the above-mentioned segment of the left lateral sinus.*

These arteriovenous malformations may also extend outside the cranium and Verbiest (1968a) separates the purely intradural AVA of the posterior fossa from those more rare ones that not only involve the dura mater but also the extracranial soft tissues and the petrosal bone.

The association of these dural malformations with other vascular lesions like intracranial arterial aneurysms has also been recorded by Verbiest (1968b), Houser *et al.* (1972), and others.

Other than the congenital origin of the dural AVM aetiological factors like trauma or local inflammatory diseases have also been considered (Aminoff, 1973). In the review of the literature and personal cases a previous history of a significant head injury with enough data on this point appeared in only 4% of 92 cases.

CLINICAL SYNDROMES

It is very important, in our view, to define properly and to separate the different clinical syndromes of these patients suffering from dural AVM of the transverse-sigmoid sinus. Previous attempts have neither been very systematic nor complete (Laine *et al.*, 1963; Nicola and Nizzoli, 1968; Verbiest, 1968; Debrun and

Chartres, 1972; Houser *et al.*, 1972; Aminoff, 1973), mainly because a large number of well documented cases are necessary for such grouping of clinical syndromes.

Several factors have to be considered in order to understand the variety of symptoms and signs:

1. Subjective disturbances like insomnia are

FIG. 2. Case 6. *Right internal carotid angiogram. The markedly enlarged lateral tentorial branch of the internal carotid artery (long arrow) drains directly into the right lateral sinus (large arrow).*

TABLE 8
ARTERIAL SUPPLY TO ARTERIOVENOUS MALFORMATIONS
OF TRANSVERSE-SIGMOID SINUS (91 CASES)

Artery	Total		Only arterial supply	
	(No.)	(%)	(No.)	(%)
External carotid	81	89	24	26
Internal carotid	47	51	2	2
Vertebral	29	31	2	2

caused by the continuous head bruit and tinnitus due to the arteriovenous shunt.

2. Cerebral hypoxia and ischaemia, also secondary to the AV shunt, are responsible for symptoms and signs of neurological deficit.

3. Increased venous pressure at the torcular region induces headaches and papilloedema. Infantile hydrocephalus with intracranial bruit is also due to a tremendous increase of venous pressure as a consequence of the massive arteriovenous shunt at the torcular region.

4. Retrograde drainage through collateral circulation sometimes produces engorgement of other basal sinuses, thus secondarily causing distant symptoms and signs.

5. Obstruction of some of the affected sinuses.

6. Involvement of the pial venous system may produce subarachnoid haemorrhages extending sometimes into the cerebral tissue leading to haematomas.

The predominance or association of several of these six main factors will no doubt provide the basis for the clinical syndrome in each particular patient. As a result of the review of the literature and our personal material we have arrived at the findings summarized in Tables 4 and 5 with symptoms and signs listed according to their frequency. Correlating the clinical signs with the six main pathophysiological factors, several groups of clinical syndromes according to their main features and pathophysiological basis have been outlined (Table 6).

It may be observed that about one-third of all patients with AVM of the transverse-sigmoid sinus have only subjective symptoms due to the bruit (group I). However, the clinical syndromes more frequently observed in this condition correspond to groups II and III or a combination of both. Altogether they form 40%

of the whole material. Besides the symptoms and signs secondary to increased venous pressure and cerebral ischaemia, in those cases there was a cranial bruit that indicated an AVF or AVM in about half of them. Subarachnoid haemorrhages represent 17% of these vascular malformations, being the first symptom in 13% of the patients (Table 4). Finally, a much less frequent syndrome is group IV with infantile hydrocephalus and cranial bruit. This was present in only 4% of all recorded patients.

Thus, there are various clinical modes of presentation of these dural arteriovenous malformations that may be confused with other neurological disorders, especially when there is no cranial bruit as a leading symptom and sign.

There were no sex differences in these lesions and the duration of the symptoms was very wide as well as the age of presentation, predominantly between 41 and 60 years (Table 7).

ANCILLARY EXAMINATIONS

Plain radiographic examination of the skull does not show important changes in most cases (Newton *et al.*, 1968). However, Aminoff (1973) found significant abnormal signs in a great number of his patients, such as prominent meningeal vascular channels, enlarged foramen spinosum, bony lacunae, and erosions.

FIG. 3. Case 6. Right external carotid angiogram. Meningeal branches of the right occipital artery (long arrow) are enlarged and drain into the right lateral sinus (large arrow).

TABLE 9
MAIN ANGIOGRAPHIC FEATURES OF AVM
OF TRANSVERSE-SIGMOID SINUS

	Cases with detailed angiographic exploration recorded in literature (46)	Personal cases (6)	Total cases (52)
Arterial supply			
Branches of occipital artery	40	6	46
Branches of middle meningeal artery	26	3	29
Tentorial branches of internal carotid artery	28	3	31
Other arteries	17	2	19
Primary venous drainage			
Transverse-sigmoid sinus	33	5	38
Cortical veins		1	1
Other veins or sinuses	7		7
Not specified	6		6
Normal anterograde drainage			
Jugular veins	5	2	7
Deep cervical veins	3		3
Retrograde drainage	13	3	16
Sinus obstruction			
Transverse-sigmoid sinus	6	4	10
Other sinuses	3		3

Radioisotopic scanning and measurement of arterial-venous oxygen saturation have also been carried out in some of these cases. Phonocardiography may be helpful for the more precise localization of the cranial bruit which is usually more intense over the temporomastoid region. But undoubtedly angiography with the recent techniques of selective arterial catheterization, subtraction, and magnification represents the only examination that may determine the extent and supply of these malformations with a greater precision.

With regard to the different branches of the external and internal carotid arteries and of the vertebral arteries which may supply the arterio-venous fistula, the findings in our cases are shown in Table 2, and Table 8 reviews the 91 cases reported in the literature, including also the personal material. Table 9 summarizes the arterial supply and venous drainage together with the retrograde drainage and sinuses obstructions of 46 cases taken from the literature with adequate angiographic data and the six personal cases. In our material we have

TABLE 10
AV MALFORMATIONS OF TRANSVERSE-SIGMOID SINUS

<i>29 cases untreated</i>	<i>Unchanged</i>	<i>Death</i>	<i>Spontaneous arrest</i>	<i>Unknown</i>	
	9	3	1 (after angiography)	16	
<i>Cases surgically treated (55; 71 operations)</i>	<i>Cured</i>	<i>Improvement</i>	<i>Unchanged</i>	<i>Death</i>	<i>Unknown</i>
Ligature of vessels at neck (40)					
Unilateral external carotid	3	4	10	2	7
Bilateral external carotid	—	1	2	3	3
Vertebral artery	—	—	4	—	—
Cervical artery	—	—	1	—	—
Direct approach (27)					
Afferent arterial ligation and removal	5	4	—	2	—
Dural incision and coagulation	6	2	—	—	—
Transverse sinus ligation	—	1	—	—	—
Unspecified	—	—	4	2	1
Combined extra-intracranial approach (4)					
Internal carotid 'trapping'	—	—	1	—	—
With carotid ligation	1	1	—	—	1

stressed the alterations in the venous drainage (retrograde drainage and obstruction of sinuses) that have not been fully considered in the literature except by Dichgans *et al.* (1972).

Several examples of the angiographic features of cases 5 and 6 (Figs 1, 2, and 3) are presented as the other four earlier cases have already been reported (Fernández Urdanibia *et al.*, 1974).

TREATMENT

Most authors have observed that occlusion and ligation of some of the afferent arteries may not be sufficient to stop the circulation of these arteriovenous malformations. The extent and multiple afferent blood supply through various channels are adverse factors for the cure or improvement of this condition. Some patients not treated or submitted only to ligation of some vessels in the neck may be improved or do not show progressive changes in their evolution (Table 10).

The direct surgical approach with occlusion or removal of these malformations has been considered the best form of treatment where this is possible (Laine *et al.*, 1963; Pecker *et al.*, 1965; Debrun and Chartres, 1972; Kosnik *et al.*, 1974). These direct attempts at removal were mainly carried out in cases with a history of subarachnoid haemorrhage and especially with signs indicating intracranial haematomas (our case 4).

Recently, Hugosson and Bergström (1974) have treated successfully some of these dural AVM by making a wide exposure of both the occipital and cerebellar dura mater followed by three long dural incisions along the occipital region to the middle cranial fossa, along the tentorium, and finally along the cerebellar dura mater following the inferior border of the transverse and sigmoid sinuses. With this technique, coagulation and obliteration of all the vascular channels and almost complete isolation of these sinuses may be obtained. Table 10 demonstrates from the recorded experience that the direct approach is much more effective. The appropriate type of treatment naturally depends on the extent and symptomatology of the vascular lesion but the tendency nowadays is towards a more direct and radical treatment. Perhaps in the future, multiple embolization by selective cath-

terization will provide the possibility for occlusion of these dural AVM. Embolization with a mixture of wax and iophendylate has also been tried in some of these lesions (P. Albert Lasiera, personal communication).

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ADDENDUM

Since this paper was written two articles have appeared: (1) Padalko, P. I., and Serbinenko, F. A. (1974. Neurosurgical pathology of the cerebral blood vessels. Moscow (in Russian)) present 32 cases of AVM of the transverse-sigmoid sinus studied at the Burdenko Institute; they emphasize that these patients are often considered to be suffering from brain tumours or vascular lesions. (2) Kosnik, E. J., Hunt, W. E., and Miller, C. A. (1974. Dural arteriovenous malformations. *Journal of Neurosurgery*, **40**, 322–329,) review this subject and report two cases of transverse-sigmoid AVM.

APPENDIX TABLE
SUMMARY OF RECORDED CASES

Authors	Sex	Age (yr)	History duration	Main symptoms	Clinical signs	Angiography		Treatment	Evolution
						Afferent supply	Venous drainage		
Tönnis (1936) Röttgen (1937)			2 cases 1 case	} Cited by Verbiest (1951)					
Verbiest (1951)	M	54	6 m after head injury		Bruit, tinnitus	Papilloedema, communica- ting hydro- cephalus	No visualized angiography		Combined tentorial and cere- bellar AVA
Obrador and Urquiza (1951, 1952)	M	20 m	17 m	Exophthalmos, vomiting, seizures instability, bruit	Macrocephaly, dilated scalp veins, bruit, exo- phthalmos	Bilat. ext. carotid arteries	Bilat. trans. sinus	Bilat. ext. carotid ligature	Died after 6 m
Fontaine <i>et al.</i> (1957)	F	56	6 m after head injury	Tinnitus, bruit	Papilloedema, bruit	Ext. and int. carotid arteries	Trans. sinus and jugular vein	Jugular and ext. carotid ligation and division	Recovered after 9 m
Ciminello and Sachs (1962)	M	58	3 h	Headache, subarach- noid haemor- rhages	Meningeal signs, dysphasia	Ext. carotid and occipital arteries	Galen's vein and torcular	Suboccipital craniotomy. Posterior meningeal artery occlusion	Recovered
Epstein and Platt (1962)	F	11 w	5 w	Cyanosis, bruit, dyspnoea	Bruit, heart failure	Visualized in angiocardio- graphy		—	Died from heart failure
Verbiest (1962)	F	40	3 yr	Bruit	—	Ext. carotid (occipital artery)	Transverse sinus	Ext. carotid and occi- pital arteries ligation	Bruit improve- ment
Laine <i>et al.</i> (1963)	M	23	2 yr	Mental re- tardation. 3 recurrent episodes of speech dis- turbances and hemi- paresis	Hemiparesis, speech disturbances	Vertebral artery	Cavernous sinus	No findings in posterior fossa and fronto- temporal craniotomy	Unchanged
—	M	33	2 yr	2 subarach- noid haemor- rhages, headaches, vertigo, visual failure	Meningeal signs. III paresis	Int. and ext. carotid and vertebral artery	Torcular and pterygoid vein	Temporal craniotomy and tentorial vessel coagulation	Recovered after 2 m
van der Werf (1964)	F	3	—	Hydrocephalus	Bruit, hydro- cephalus, papilloedema	Bilat. int. and ext. carotid and vertebral arteries	Transverse sinus and torcular	Bilat. dural incision from tem- poral fossa to free edge of tentori- um. Bilat. ext. carotid ligation	Bruit disap- peared
Pecker <i>et al.</i> (1965)	F	56	15 yr	Bruit, head- aches, cer- vical pain, vertigo	Bruit	Ext. carotid artery	Transverse sinus and jugular vein	Ext. carotid ligation	No change after 7 yr
—	M	36	Brief period	3 subarachnoid haemor- rhages Episodes	Papilloedema bilateral VI palsy. Meningeal syndrome	Ext. carotid and vertebral arteries	Anomalous vein crossing cisterna magna	Ext. carotid ligation and suboccipital craniotomy with ligation of venous drainage	Recovered
Takekawa and Holman (1965)	F	49	1 yr	Headaches and bruit	Bruit	Ext. carotid artery	Transverse sigmoid sinus	Ext. carotid ligation	Recovered
	M	28	5 m	Proptosis ocular pain	Bruit, proptosis	Ext. carotid artery	Jugular vein	Ext. carotid ligation	Recurrence of chemosis after 2 m

APPENDIX TABLE—continued

Authors	Sex	Age (yr)	History duration	Main symptoms	Clinical signs	Angiography		Treatment	Evolution
						Afferent supply	Venous drainage		
van Wijngaarden and Vinken (1966)	F	65	5 m	Bruit	Bruit	Ext. carotid artery	Transverse sinus	Occipital artery ligation and ext. carotid ligation 1 m later	Bruit improvement
Newton <i>et al.</i> (1968)	M	22	—	Headaches	Bruit, papilloedema	Int. carotid, bilat. ext. carotid, vertebral artery	Galen's vein	Bilat. ext. carotid ligation and ventricular shunt	—
	M	1	—	Macrocephaly	Bruit, dilated scalp veins	Int. carotid and bilat. ext. carotid arteries	Torcular	Ventricular shunt and ligation of bilat. ext. carotid and tentorial arteries	—
	M	33	5 yr after head injury	Bruit, retroauricular mass	Retroauricular mass, bruit	Bilat. int. carotid, ext. carotid and vertebral arteries	Transverse sinus	Ext. carotid ligation and removal of extracranial mass	—
	F	3½	Brief period	Vomiting, gait disturbance	Hemiparesis, proptosis	Bilat. ext. carotid, int. carotid and vertebral arteries	Straight sinus and dural lake	—	—
	M	47	2 yr	Exophthalmos, seizures, arterial hypertension	Exophthalmos, hemianopsia	Int. and ext. carotid arteries	Straight sinus	—	—
	M	51	—	Subarachnoid haemorrhage	Meningeal signs	Trigeminal artery, int. and ext. carotid arteries	Transverse and superior sinus	Occipital angioma and haematoma removal	—
	M	50	—	Hemiparesis	VII palsy, and dysaesthesia	Int. carotid artery	Straight sinus, basal vein, and Galen's vein	Common carotid ligation and ligation of meningeal branches intracranially	—
	F	43	—	Bruit, occipital pain	Bruit	Int. and ext. carotid arteries	Transverse sinus	—	—
	F	29	—	Bruit	Retroauricular mass. Bruit, dilated scalp veins	Int. and ext. carotid arteries	Transverse sinus and superficial veins and superior longitud. sinus	—	—
	M	68	—	Visual failure, ocular pain	Bruit, papilloedema, visual failure	Int. carotid and bilat. ext. carotid arteries	Transverse sinus	—	—
	F	49	—	Visual failure, bruit, arterial hypertension	Bruit, papilloedema, blindness in R eye	Bilat. int. carotid. Ext. carotid artery	Transverse sinus	Occipital artery ligation and occipital craniotomy	—
	M	52	—	Transient ischaemic episodes	—	Vertebral artery	Superficial cerebellar vein	—	—
	F	58	1 yr	Bruit	Bruit	Ext. carotid artery	Transverse sinus	—	—
	M	52	3 yr	Bruit	Bruit, loss of hearing	Ext. carotid artery	Transverse sinus	—	—
	F	66	4 m	Bruit, arterial hypertension	Bruit, nystagmus	Ext. carotid artery	Transverse sinus	—	—

APPENDIX TABLE—continued

[illegible]

APPENDIX TABLE—continued

Authors	Sex	Age (yr)	History duration	Main symptoms	Clinical signs	Angiography		Treatment	Evolution
						Afferent supply	Venous drainage		
Legré <i>et al.</i> (1969)	F	27	14 yr	Seizures, headaches, bruit	Bruit	Bilat. vertebral artery. Ext. carotid	Transverse and sigmoid sinus	Occipital and ext. carotid ligation. 3 m later ligation of vertebral artery branches. 3 m later vertebral ligation	Bruit recurrence after 3 yr
	M	46	8 yr	Headaches, bruit	Bruit, facial hypaesthesia	Ext. carotid and vertebral arteries	Transverse sinus and jugular vein	Ligation of occipital and branches of vertebral artery	Bruit improvement
	M	59	3 m	Transitory aphasia, headaches, subarachnoid haemorrhages, bruit	Normal	Ext. and int. carotid and vertebral arteries	Transverse sinus and cortical drainage to sup. longitud. sinus	—	Unchanged
	F	60	1 yr	Bruit	Bruit	Ext. carotid artery	Transverse sinus	Occipital artery ligation and excision of malformation	Bruit improvement
Decker and Backmund (1970)	M	3 m	Days	Subarachnoid haemorrhage	Meningeal syndrome	Ext. carotid artery	Jugular vein	—	—
Robinson and Sedzimir (1970)	M	19 m	—	Seizures, hemiparesis—subcutaneous angioma	Bruit, hemiparesis	Ext. carotid and vertebral arteries	Transverse sinus and venous lake	External carotid resection 1 m later suboccipital craniotomy. Later ventricular shunt	Died
Billewicz <i>et al.</i> (1971)	M	3½	2 m	Drowsiness, headaches, vomiting, speech and gait disturbance	Bruit, papilloedema hemiparesis, cerebellar syndrome	Ext. carotid artery	Transverse and sigmoid sinus. Retrograde drainage to straight sinus and sup. longitud.	Ext. carotid ligation	Partial improvement after 2½ yr
Debrun and Chartres (1972)	M	6 m	—	Dilated scalp veins. Occipital angioma, convulsive seizures	Bruit, psychomotor retardation	Bilat. ext. and int. carotid arteries, vertebral artery	Bilat. transverse sinus and sup. longitud. sinus	Bilat. ext. carotid ligation	Bruit recurrence 3 m later
	F	47	—	Subarachnoid haemorrhages, headaches	Meningeal syndrome	Int. carotid artery	Bilat. transverse sinus and torcular	—	Unchanged
Dichgans <i>et al.</i> (1972)	F	64	m	Tinnitus, bruit	Bruit	Ext. carotid artery	Transverse sinus, sigmoid sinus occluded	—	—
	F	72	6 m	Bruit, tinnitus	Bruit	Ext. and int. carotid arteries	Cortical veins and superficial petrous sinus	—	—

APPENDIX TABLE—continued

Authors	Sex	Age (yr)	History duration	Main symptoms	Clinical signs	Angiography		Treatment	Evolution
						Afferent supply	Venous drainage		
	M	41	Hours	Subarachnoid haemorrhage	Bruit, meningeal signs, coma	Ext. and int. carotid arteries, middle cerebral artery	Transverse sinus and cortical veins to the superior longitudinal sinus. Transverse sinus occluded	—	—
	F	55	2 m	Headaches, tinnitus, seizures	Papilloedema	Ext. and int. carotid arteries	Sup. longit. and transverse sinus thrombosed. Retrograde cortical venous drainage	—	Exophthalmos and seizures 10 yr later
	M	55	2 yr	Seizures, loss of vision, bruit, loss of hearing	Bruit, papilloedema	Ext. carotid artery	Transverse sinus and jugular vein. Retrograde drainage to straight and cavernous sinuses	—	—
	—	70	1 m	Subarachnoid haemorrhage, left parietal syndrome, bruit	Bruit, papilloedema, hemianopsia, hemiparesis	Ext. and int. carotid arteries	Transverse sinus and retrograde cortical vein drainage. Sigmoid sinus occluded	—	—
Houser <i>et al.</i> (1972) (12 occipital cases)	F: 9 M: 3	20–29 (1) 30–39 (1) 40–49 (1) 50–59 (1) 60–69 (1) 70 (1)	< 1 yr	Bruit (10), seizures (2), headaches (7), diplopia (1), proptosis (1), visual failure (1), subarachnoid haemorrhages (3), intracerebral hrge (1), subdural hrge (1)	Bruit (10), cerebellar syndrome (3), proptosis (1), visual failure (1), ocular palsy (1), cerebral deficit (1), pontocerebellar deficit (1)	Occipital artery (9), middle men. artery (9), meningeal hypophyseal artery (5), vertebral artery (2), ascending pharyngeal artery (2)	—	Surgical (5), ext. carotid ligation (5), direct approach (3)→Unchanged (3) radiotherapy (1)→Complications (1) Non-treated (7)	
(3 posterior fossa cases)	M M M	20–29 (1) 30–39 (1) 40–49 (1)							
			12 yr–1	Headaches (1), seizures (1), ocular pain (1), proptosis and subarachnoid haemorrhage (1)	Bruit (2), proptosis, chemosis (1)	—	—	Surgical (2), ext. carotid ligation (1), direct approach (1), untreated (1)	Recovered (1) complications (1)
Senarclens <i>et al.</i> (1972)	M	52	—	Tinnitus, bruit, headaches	Bruit	Ext. carotid artery	Sigmoid sinus and jugular vein	—	Unchanged
	F	58	—	Headaches, bruit, 2 subarachnoid haemorrhages	Bruit, meningeal signs	Bilat. ext. carotid arteries, vertebral artery	Torcular	Bilat. occipital ext. carotid artery ligation	Died after 4 yr subarachnoid haemorrhage
	M	41	—	Tinnitus, loss of hearing, headaches, mental deterioration, aphasia	Drowsiness, papilloedema hemianopia, dysphasia	Ext. and int. carotid arteries	Transverse sinus and retrograde drainage to straight sinus and cortical veins, sigmoid sinus occluded	Ext. carotid ligation	Unchanged 2 m later

APPENDIX TABLE—continued

Authors	Sex	Age (yr)	History duration	Main symptoms	Clinical signs	Angiography		Treatment	Evolution
						Afferent supply	Venous drainage		
Aminoff, 1973	M	38	36 h	Headaches, visual failure, sub-arachnoid haemorrhage	Bruit, hemianopsia	Ext. and int. carotid arteries	Transverse sinus, sigmoid sinus, and superior longitudinal sinus	Excision	Loss of vision 2 yr later
	M	53	5 yr	Headaches, visual failure, sub-arachnoid haemorrhages	Bruit, papilloedema, blood in csf	Bilat. ext., int. and vertebral arteries	Torcular	Craniotomy, occlusion of feeding vessels	Recovered after 6 m
	F	43	1 yr	Tinnitus, headaches	Bruit	Ext. carotid artery	Transverse sinus	—	Unchanged
	F	58	6 w	Episode of loss of consciousness, visual hallucinations	Bruit	Ext. carotid artery	Transverse and straight sinuses	—	Unchanged
	F	62	2 d	Speech disturbances, subarachnoid haemorrhage	Dysphasia, bilateral Babinski signs, blood in csf	Ext. and int. carotid arteries	Venous sinus drainage to transverse sinus	Excision	Focal seizures 10 y later
	M	42	3 m	Tinnitus	Bruit	Ext. and int. carotid arteries	Sigmoid sinus	—	Unchanged 2 m later
	M	54	12 yr	Headaches, seizures, mental deterioration, visual failure	Bruit, exophthalmos, hemiparesis ataxia, brain stem syndrome	Bilat. ext., int., and vertebral arteries	Bilat. transverse sinus	Craniotomy and excision of feeding vessels	Partial improvement 3 yr later
Aminoff and Kendall	M	48	2 yr	Headaches, loss of vision (ventricular shunt due to papilloedema 9 m earlier)	Bruit, optic atrophy	Ext. and int. carotid arteries	Transverse and sigmoid sinus, temporal superficial veins and cervical veins	—	Died, thrombosed cerebral veins
(Associated with other intracranial pathology)	M	69	—	—	—	Ext. and int. carotid arteries	Transverse sinus	—	Died (cerebral metastasis from lung cancer)
	F	50	—	—	—	Ext. and int. carotid arteries	Sigmoid sinus	Removal of frontal tumour	Died (frontal astrocytoma)
	F	65	—	—	—	Ext. carotid artery	Superficial vein drainage to superior longitudinal	—	Died (tuberculous meningitis)
	F	59	—	—	—	Ext. and int. carotid arteries	Transverse sinus	Removal of frontal tumour, 6 m later occlusion of malformation afferent veins, ventricular shunt	Improving slowly (frontal meningioma)
Storrs and King (1973)	M	50	1 yr	Tinnitus, headaches, loss of vision	Bruit	Ext. and int. carotid, and vertebral arteries	Sigmoid sinus and jugular vein	Embolization, ext. carotid ligation, excision of malformation	Recovered after 2 yr

APPENDIX TABLE—continued

Authors	Sex	Age (yr)	History duration	Main symptoms	Clinical signs	Angiography		Treatment	Evolution
						Afferent supply	Venous drainage		
Hugosson and Bergström (1974)	M	36	6 m	Tinnitus, headache	Bruit	Ext. and int. carotid vertebral arteries	—	—	Spontaneous arrest after angiography, recovered after 1 yr
	F	48	—	Bruit	Bruit	Ext. carotid artery	Transverse and sigmoid sinuses	Dural section above and below transverse sinus and occlusion of afferent veins	Recovered
	M	24	—	Bruit	Bruit	Ext. and int. carotid and vertebral arteries	Transverse and sigmoid sinuses	Dural section above transverse sinus, ligation of branches of ext. carotid artery	Improvement of bruit
	M	52	—	Bruit	Bruit	Ext. and int. carotid and vertebral arteries	Transverse and sigmoid sinuses	Dural section above and below transverse sinus and occipital dura mater	Recovered
	M	39	—	Bruit	Bruit	Ext. and int. carotid arteries	Transverse and sigmoid sinuses	Dural section above and below transverse sinus, occlusion of superior petrous sinus. Ligation ascending pharyngeal artery	Improvement of bruit
Kosnik <i>et al.</i> (1974)	M	71	—	Bruit	Bruit	Ext. and int. carotid and vertebral arteries	Transverse and sigmoid sinuses	Occipital artery ligation	Unchanged
	F	49	2 m	Bruit	Bruit	Ext. carotid and vertebral arteries	Sigmoid sinus and jugular vein	Ext. carotid artery resected, 5 m later vertebral artery ligation, suboccipital craniotomy, coagulation of dural veins	Recovered
	F	52	6 m	Vomiting, headaches, subarachnoid haemorrhage, neurological deterioration	Bruit, loss of vision, papilloedema, ataxia, hemianopsia	Ext. and int. carotid and vertebral arteries	Torcular, superior longitudinal sinus, cortical veins	Bilat. ext. carotid ligation, craniotomy with partial coagulation of the angioma, 1 yr later ligation of feeding arteries and transverse sinus ligation, 9 m later ligation of longitud., transverse and straight sinuses, excision around torcular, tentorium and falx	Bruit disappeared

Six personal cases, see Tables 1, 2, and 3.