Angiography of Pulseless Disease
A Study of 50 Cases

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脈なし病50例の血管撮影

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大動脈弓症候群を呈する50例のいわゆる脈なし病症例につき血管造影所見を分析し検討を加えた．Seldinger 氏法を用い胸部並びに腹部大動脈撮影を全例に，分枝の選択的撮影を17例に施行した．大動脈弓主幹動脈の閉塞性病変は全例に証明されたが，これらに動脈瘤を併有するものが2例ある．病変は左鎖骨下動脈（41例），左総頸動脈（30例），右鎖骨下動脈（26例）の順に多いが，左では近位鎖骨下動脈病変（椎骨動脈分岐点以前）が多く，右では遠位鎖骨大動脈病変の多いのが特徴である．腎動脈狭窄は32%に認められ非狭窄群に比して高血圧の頻度が明らかに高いが同時に存在する大動脈狭窄と大動脈弁閉鎖不全による寄与を考慮すべきである．腹部大動脈前方枝の中では上腸管膜動脈の閉塞が目立つ（9例）．左冠動脈起始部に動脈瘤形成を示すものが1例ある．大動脈本幹の異常は全症例の64%に認められ，狭窄（25例），拡張（12例），大動脈瘤（2例），壁不整化（23例），壁肥厚（12例），屈曲蛇行（7例）等であり，更に大動脈弁閉鎖不全を4例に認める．これ等の大動脈病変に注目して全症例を，(1) 大動脈の狭窄を主徴とする群（46%），(2) 狭窄以外の諸変化を主徴とする群（18%），(3) 弓部及びその分枝に病変が限られる群（36%）の3群に分類出来る．腹部大動脈領域には約40%の症例が何等かの異常所見を示すので腹部大動脈撮影が必須となる．側副路の発達は全般に極めて良好であるが，連続フイルム上に現れた全ての側副路を整理分類した．動脈硬化性病変で注目されているいわゆる "Subclavian steal" の頻度は低く（7例），鎖骨下動脈閉塞に対する側副路としては肋間動脈によるもの（19例），甲状頸動脈の分枝によるもの（27例）の方が強力である．上胸部側副吻合網と名づけた特異な側副路は大動脈弓主幹動脈に広範な閉塞を有する症例に特徴的である．腹部大動脈の狭窄やその前方枝の閉塞では11例に meandering mesenteric artery による側副血行を認めた．

Introduction

In 1908 Takayasu[35], a Japanese opthalmologist, reported a peculiar arteriovenous anastomosis of the retinal vessels in a young female patient. It is generally agreed that this is the first documented case of pulseless disease (Takayasu's disease). The eye sign reported by Takayasu is undoubtedly the manifestation of inadequate carotid circulation. However, Takayasu did not describe pulsations of the arm and neck. It was Ohnishi and Kagoshima who reported similar cases and stressed that absence of the radial pulses was another important feature of the disease[35]. Shimizu and Sano[34], Japanese surgeons, first used

the term "pulseless diseass" to describe this condition in 1948. They consider pulseless disease to be a chronic obliterative bracheocephalic arteritis of unknown etiology, causing various ischemic signs in the distal parts of the occluded vessels. Case reports of this disease have been seen predominantly in Japan until in 1954 Ask-Upmark[2] Published a paper "On the pulseless disease outside of Japan". Since then it has been known that this condition is not infrequently seen also in Western countries. The term "Aortic arch syndrome", first suggested by Frøvig,[16] was more clearly defined by Ross and Macusick as diminished or absent pulses in arteries arising from the arch of the aotra regardless of cause.[23,32] Ask-Upmark[3] described pulseless disease as a form of this clinical syndrome.

Thus, the pathological changes of pulseless disease have been considered to be mainly in the aortic arch and its branches. It was not until 1959 when Danaraja[12] reported cases with arteritis of a similar type in the abdominal region, that attention began to be paid to the entire aorta and its branches. Inada[21] in 1961 reported 6 cases of pulseless disease with coarctation of the abdominal aorta. Nasu[28] in 1963 also showed in his excellent review of autopsy cases that the changes are not necessarily limited to the aortic arch but spread to the thoracic aorta, abdominal aorta and even to the main pulmonary arteries. Recently Ueda[40] has suggested a more generalized term "Aortitis syndrome" to describe the widespread abnormalities seen in this condition. Regardless of confusion in terminology, reports have been seen with increasing frequency world wide[18,19,24,37]. The etiology of this disease is still unknown.

Selective angiography of the aorta and its branches is the best diagnostic procedure known at present time. The exact location and degree of abnormalities are clearly visualized and deffinite informations needed for treatment of the patient is obtained. The purpose of this study is to present angiographic findings in 50 cases of pulseless disease with discussion and review of the literature.

Material

During the period of July, 1963 to October, 1966, 69 cases of aortic arch syndrome underwent angiographic study in the Department of Radiology, Tokyo University. Of these, 59 cases were the so-called pulseless disease or Takayasu arteritis type (Table 1). Of these, 9 were excuded; 4 cases in which films

Table 1. Classification of 69 cases of aortic arch syndrome

Pulseless disease	59
Atherosclerosis	3
Thromboangitis obliterans	2
Syphilitic aortitis	1
Dissecting aneurysm	1
Congenital ?	1
Traumatic?	1
Thoracic outlet compression syndrome	1

were not available (still in use in another department), 1 cases with previous surgical intervention (resection of aneurysm) and 4 cases in which examination of the abdominal aorta was not done. Thus, a total of 50 cases were chosen for the analysis. The age distribution ranged from 18 to 50 with the peak incidence in the third decade. (Fig. 1). All these cases showed a history of absent or weak pulses in the neck or arm of about 4 years' average duration. Serologic tests for syphilis were negative and serum cholesterol level was within the normal range. Acceleration of blood sedimentation rate, strongly psotiive C-reactive protein reaction and increased serum gamma-globulins were frequently observed. Typical changes of the

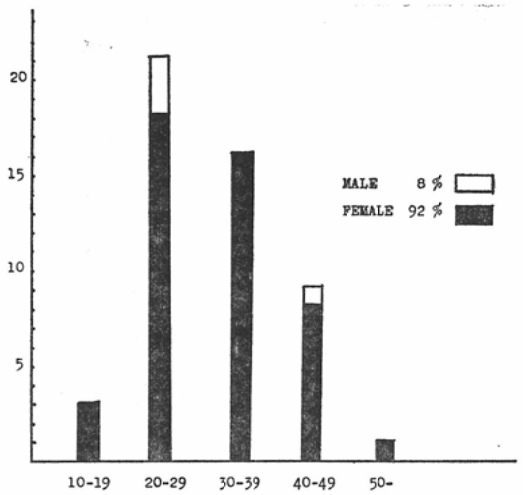


Fig. 1 Age and Sex distribution

central retinal vessels as described by Takayasu were encountered in only 6 cases.

Method

Percutaneous thoracic and abdominal aortography were perfoemed in all 50 cases; in addition, in 17 of these one or more selective injections into the branches of the aorta were made at the same sitting (Table 2). The catheter was introdeced usually via the right femoral artery and the trans-axillary route

Table 2. Examinations performed in 50 cases of pulseless disease.

Thoracic aortography	all cases
Abdominal aortography	all cases
Selective arteriography	17 cases
Innominate artery	12 cases
Left subclavian artery	10
Left common carotid artery	3
Right common carotid artery	2
Left vertebral artery	1
Celiac artery	1
Both renal arteries	1

was used in only one case with complete obstruction of the abdominal aorta. Under fluoroscopic guidance the catheter tip was first positioned about 5 cm. above the aortic valve and 40 to 45 cc. of 76% Urografin or 80% Angio-conray was injected under a pressure of 10 kg/cm^2. Then, the catheter was withdrawn to the abdominal aorta and the tip was positined at the level of the lower margin of the first lumbar vertebra and 30 to 35 cc. of the contrast medium was injected under a pressure of 7 kg/cm. For selective injection 60% Urografin or Conray was used. Serial roentgenograms were made for 3 or 5 seconds after the injection.

Angiographic Findings

I). Changes in the Branches of the Thoracic and Abdominal Aorta

As indicated in Table 3, occlusive changes (aneurysmal dilatation, rarely) were seen in all the main

Table 3. Frequency of Branch Lesions in 50 Cases

Brachiocephalic branches	50 (100%)
Renal arteries	16 (32%)
Celiac artery	1 (2%)
Superior mesenteric artery	9 (18%)
Inferior mesenteric artery	1 (2%)
Intercostal arteries	18 (36%)
Lumbar arteries	6 (12%)
Coronary arteries	1 (2%)

Table 4. Involvement of Bracheocepharic Branches in 50 Cases

	complete occlusion	Stenosis severe	Stenosis moderate	Stenosis mild	Aneurysmal dilatation	Total	Per Cent (Total/50) cases
Innominate artery	4	3	3	3	0	13	26%
Rt. common carotid artery	3	4	3	5	0	15	30%
Rt. subclavian artery	16(3)	7(0)	2(0)	0	1(0)	26(3)	52%
Rt. vertebral artery	0	1	2	3	0	6	12%
Lt. common carotid artery	12	3	7	8	1	30	60%
Lt. subclavian artery	27(20)	6(3)	5(3)	3(3)	0	41(29)	82%
Lt. vertebral artery	0	1	1	4	0	6	12%
Total	62	24	23	26	2	137	

( ): Subclavian lesion proximal to the origin of vertebral artery

Table 5. Roentgenographic appearances of bracheocephalic lesions.

Complete occlusions		Stenosis		Aneurysm	
Flame-shaped	18	Irregular diffuse	30	Irregular fusiform	2
Immediate transition to collateral vessels	14	Smooth diffuse	32		
Cut off	8	Localized	11		
Not clear	22				

branches of the aorta.

(a) Bracheocephalic arteries

The location and degree of bracheocephalic lesions are summerized in Table 4. There were 137 lesions among 50 patients. Of these, 135 were occlusive changes and 2 were aneurysmal dilatations. The left subclavian artery was most frequently affected, left common carotid and right subclavian being the next in this order. However, the incidence of multiple lesions was frequent, occuring in 41 cases (82%). In the left subclavian artery, 90% of the lesions were located in the segment proximal to the origin of the vertebral artery, while in the right subclavian artery 70% were located distal to this point. Severe stenosis of the subclavian artery extended into the axillary artery in 4 cases. In the common carotid artery, the lesions were located in the proximal portion near the origin and the bifurcation was rarely involved. Of the 135 occlusive changes, 62 were complete obstruction and 73 were incomplete obstruction. As indicated in Table 5, the appearance of these lesions were variable. In complete occlusion, flame-shaped interruption of the vessels was most frequent. (Fig. 2). An occluded subclavian

Fig. 2. a) Occlusion of the left subclavian artery-demonstrated by thoracic aortogram.

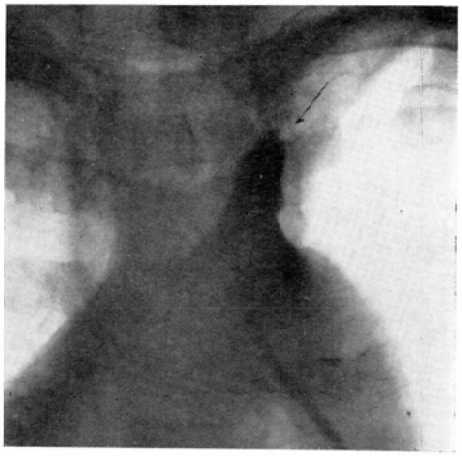


Fig. 2. b) Flame-shaped appearance of the occluded vessel more clearly demontrated by selective injection. (The same case as in a)

Fig. 3. a) Occlusion of the right subclavian artery immediately beyond the origin of the vertobral artery. A well dilated transvers scapular artery is the main collateral route. Selective injection into the innominate artery (0.5 sec).

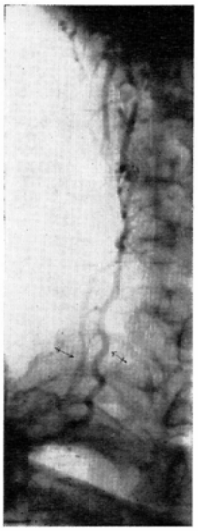


Fig. 3. b) Collateral flow comes also from the "cervical collateral network" via the deep cervical and ascending cervical artery (1.0 sec.)

artery was often directly connected to a collateral vessel which was greatly dilated approaching the same size as the subclavian. Among incomplete obstructions, a smooth and diffuse (more than 1 cm. in length) stenosis was the most frequent type (Fig. 4). However, a diffuse irregular stenosis was seen with almost equal frequency. Localized stenosis (not exceeding 1 cm. in length) was infrequent (less than 15%).

(b) Renal artery

With abdominal aortography, renal arteries were clearly visualized in all cases and renal artery

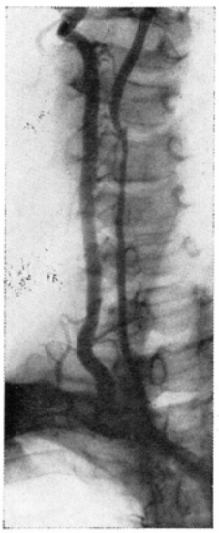


Fig. 4. a) A smooth stenosis of the right common carotid artery from the origin to the bifurcation. The external carotid artery is occluded. Note enlargenent of the vertebral artery.

Fig. 4. b) The same case as in a). Aneurysmal dilatation of the left common carotid artery at its origin.

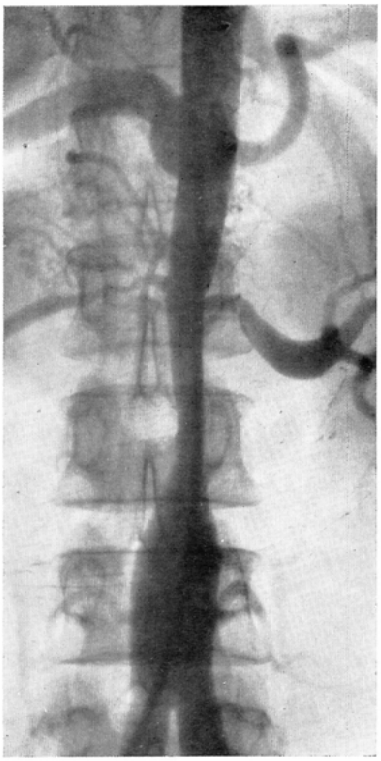


Fig. 5. Bilateral stenosis of renal arteries associated with stenosis of the abdominal aorta. The third lumbar arteries not visualized on both sides.

stenosis was demonstrated in 16 of them (Fig. 5, Table 6). The stenosis was bilateral in 5 cases and unilateral in 11 cases. The criteria of stenosis was "diminution of more than one third of the internal caliber. Renal artery stenosis was always associated with some abnormality of the aorta; stenosis (10), aneurysm (1), notching of the wall (2), dilatation with notching (3). In 31 out of 50 cases, a complete

Table 6. Renal artery stenosis in 16 cases

Case No.	Stenosis		Maximum blood pressure (mm.Hg.)	
	rt.	lt.	Upper extremity	Lower extremity
1	+	+	154/126	unobtainable
2		++	112/86	not recorded
3	++	+	168/55	150/-
4	++	+	170/70	204/82
5		+	140/100	138/72
8	+		162/52	154/52
15	+		206/50	210/84
19	+		134/0	260/0
20	+		158/92	not recorded
23	++	++	unobtainable	182/80
27	+		unobtainable	138/-
37		++	210/90	220/100
42	+		190/82	not recorded
44		+	172/78	168/80
47		++	188/94	188/112
50	++	++	158/64	160/66

+: mild stenosis

++: severe stenosis

Table 7. Incidence of Hypertension

	Upper extremity	Lower Extremity
Group with Renal Artery	90% (9/10)	50% (5/10)
Group without Renal Artery Stenosis	24 (5/21)	19 (4/21)
Total	45 (14/31)	29 (9/31)

*A blood pressure of more than 150/90 in upper extremity or 180/100 in lower extremity was considered as hypertension.

record of blood pressure was available both in upper and lower extremities. The incidence of hypertension was definitely higher in cases with renal artery stenosis as indicated in Table 7. However, as shown in Fig. 6, most of the hypertensive cases also had stenosis of the aorta or aortic incompetence in association with renal artery stenosis. In general, systolic pressure was elevated while diastolic pressure remained normal or low.

(c) Anterior visceral branches of the abdominal aorta

Celiac artery, superior and inferior mesenteric artery originate from the anterior aspect of the abdominal aorta. Therefore it was not easy to detect the pathologic changes of these arteries on routine anteroposterior aortograms. However, in a presence of a well developed collateral circulation mainly through the "meandering mesenteric artery", occlusive processes of one of these arteries could be diagnosed with reasonable accuracy on the serial roentgenograms. Involvement of the superior mesenteric artery was by far the most frequent and observed in 9 cases. The celiac artery and the inferior mesenteric artery were affected in 1 case respectively. Among these, there was no case in which definite symptoms referable to abdominal angina except for 1 case with a history of vague abdominal pain and diarrhea improved by by-pass operation (Fig. 12).

(d) Intercostal and lumbar artery

Opacification of the intercostal artery was absent or incomplete in 18 cases. Of these, 14 cases were associated with abnormal findings in the thoracic aorta, while in 4 cases the aorta was normal. Occlusive change of the lumbar artery was seen in 6 cases and all of them had stenosis or notching of the aorta (Fig. 5).

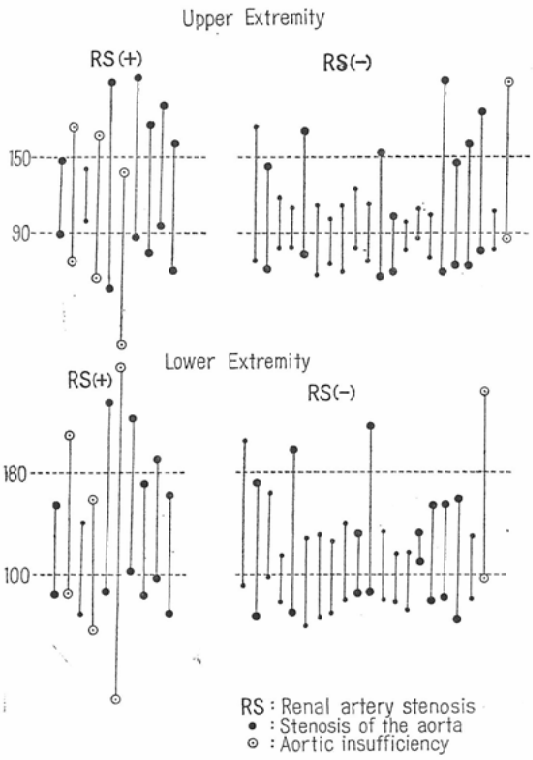


Fig. 6. Relations between blood pressure and renal artery stenosis, aortic stenosis or insufficiency.

(e) Coronary artery

Satisfactory opacification of bilateral coronary arteries was obtained in 12 cases. The left coronary artery was not visualized in 25 cases, and the right coronary in 12 cases. As no particular effort was made to opacify the coronary system, non-visualization of the coronary artery on the aortogram did not necessarily mean coronary occlusion. Aneurysmal dilatation of the origin of the left coronary artery was clearly demonstrated in 1 case with episodes of precordial pain and discomfort for several years. Electrocardiogram of this case showed no definite abnormality.

II) Abnormalities of the Aorta

Various types and incidence of angiographic abnormalities of the aorta are summerized in Table 8, and all cases of this series are classified into three major groups as shown in Fig. 7.

(a) Stenosis

Stenosis of the lumen was found in any protion of the aorta except the asceding aorta. Twenty five cases had stenosis with more than one-third diminution of the internal caliber of the aorta compared with that of the immediately proximal or distal segment. Three cases showed stenosis in the thoracic and abdominal aorta with a normal segment in between. Thus, total sites of stenosis observed were 28. In one

Table 8. Angiographic Abnormalities of the Aorta

Stenosis	25 Cases	(50%)
Dilatation	12	(44)
Aneurysm	2	(4)
Notching of the wall	23	(46)
Thickening of the wall	12	(24)
Tortuosity	7	(14)
Aortic regurgitation	4	(8)

cases a localized isthmus stenosis resembling congenital coarctation of the aorta was seen (see Fig. 7, Group 1, No. 25). Others were all so-called atypical coarctation. The incidence was 8 in the descending aorta, 7 in the abdominal aorta, 10 in that portion from the descending thoracic to the abdominal aorta. Most of them might be referred to as "elongate coarctation" and the lesions less than 5 cm. in length were only 5 in number. The wall of the affected portions of the aorta was irregularly notched in 8, while minor irregularities were noted in 20.

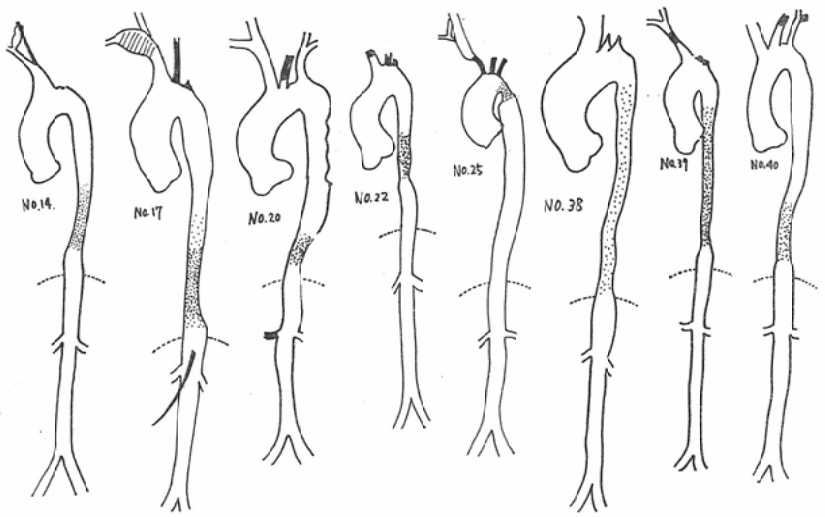
(b) Dilatation

Dilatation of the aorta (more than 40 mm. in ascending aorta and arch, more than 30 mm. in descending and abdominal aorta) was seen in 12 cases excluding 2 cases with apparent aneurysm formation. Of these, 8 cases showed the dilatation only in the ascending aorta and the arch proximal to the stenosis of the descending aorta. In these cases, the wall of the dilated segment had smooth margins, Four cases showed a diffuse dilatation of the entire thoracic aorta down to the abdominal aorta. In these cases, the wall of the affected segment had an irregularly notched appearance.

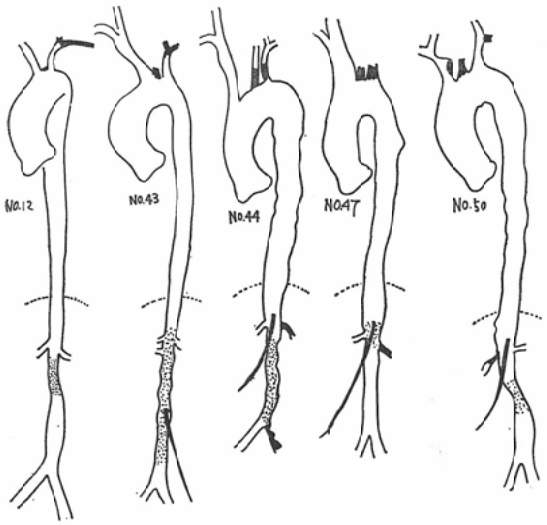
(c) Aneurysmal Dilatation

Marked aneurysmal dilatation was observed in 2 cases. One of them showed a huge saccular ane-

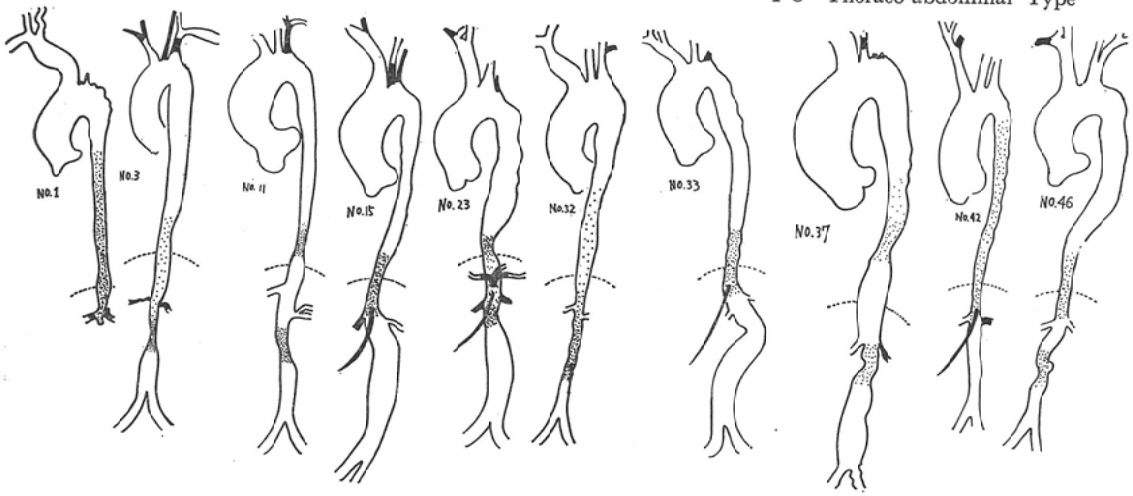
Fig. 7. Classification of 50 cases according to aortic abnormalities
Group I Stenosis of the Aorta



I-A Thoracic Type

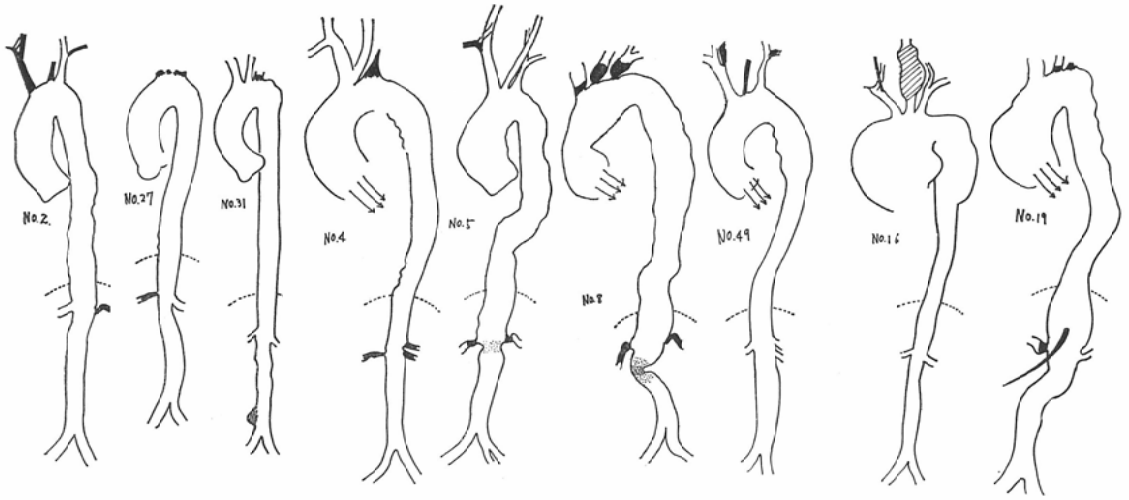


I-B Abdominal Type



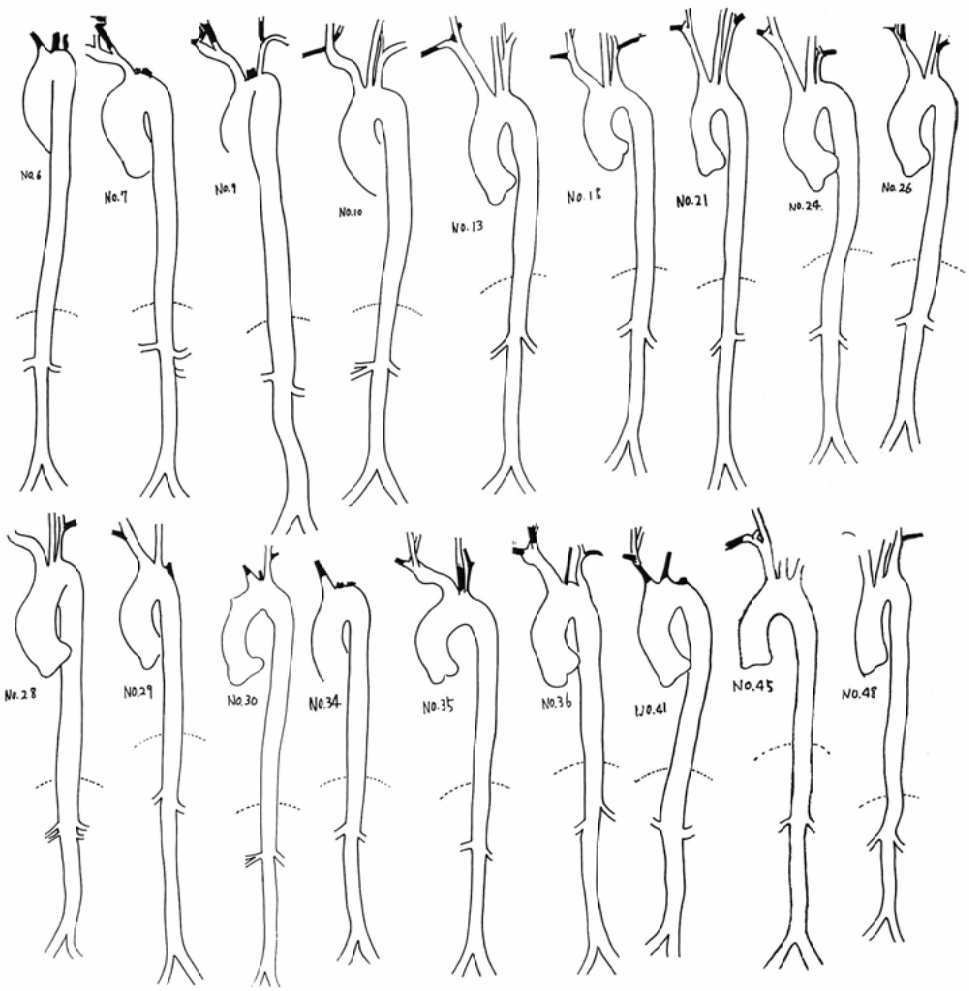
I-C Thoraco-abdominal Type

Group II Aortic Abnomalities Other Than Stenosis



II-A Notching II-B Dilatation with Notching II-C Aneurysmal Dilatation

Group III Occlusive Disease Restricted in the Major Branches of the Aortic Arch



urysm involving the ascending aorta to the proximal descending aorta. In this case a fusiforma aneurysm was also noted in the origin of the left common carotid artery. The other case showed fusiform aneurysms both in the ascending aorta and abdominal aorta.

(d) Notching of the wall

In 23 cases the inner aspect of the wall of the aorta showed an irregular notched appearance.

(e) Thickening of the wall

Marked thickening of the wall of the aorta was clearly demonstrated when the left border of the descending aorta was seen with the aid of air contrast in the left lung. (Fig. 8).

(f) Tortuosity

Marked tortuosity of the thoracic or abdominal aorta was seen in 7 cases. The course of the aorta was often more or less tortuos when stenosis or dilatation were present.

(g) Aortic regurgitation

Fig. 8. Thickening of the aortic wall demonstrated on the left side of the discending portion.

Table 9. Classification of 50 Cases According to Accompanying Abnormalities of the Aorta

Group I	Stenosis of the Aorta		
	A) Thoracic Type	8	
	B) Abdominal Type	5	
	C) Thoraco-abdominal Type	10	
		23	(46%)
Group II	Aortic Abnormalities other than Stenosis		
	A) Notching of Wall	3	
	B) Dilatation	4	
	C) Aneurysm	2	
		9	(18%)
Group III	Abnormalities Restricted Mainly in Aortic arch Branches		
		18	(36%)

Aortic regurgitation was demonstrated in 4 cases by thoracic aortography. All of these were associated with dilatation or aneurysm of the ascending aorta. One patient died six months after the examination and autopsy revealed marked dilatation of the aorta due to a severe inflammatory process. However, no definite abnormality of the aortic valve was present except for slight thickenning.

III) Collateral Circulation

Classification and incidence of various collateral patterns are summerized as shown in Table 10.

(1) Collaterals of Bracheocephalic Occlusion

Development of the collateral vessels was most remarkable in the cervical and thoracic region in the cases with occlusive lesions of the bracheocephalic arteries. Eighty eight per cent of these cases showed one or more patterns of the following collateral pathways.

(a) Transverse scapular and superficial cervical pathway

A thick dilated transverse scapular artery, sometimes in association with superficial cervical artery,

Table 10. Classification of Collateral Routes

I)	CERVICO-THORACIC REGION	
	1) Transvers scapular and superficial cervical pathway	27
	2) Reversed vertebral flow (Subclavian steal)	7
	3) Aortic intercostal pathway	19
	4) Anterior intercostal pathway	5
	5) Inferior thyroid pathway	7
	6) Pathway through the "cervical collateral network"	14
	7) Pathway through the "superior thoracic collateral network"	16
II)	ABDOMINAL REGION	
	A) Celiac or mesenteric occlusion	
	1) Pathway through meandering mesenteric artery	9
	2) Pathway through the "pancreatico-duodenal arcade"	3
	3) Pathway through the "Buhler's artery"	1
	B) Renal artery stenosis	
	1) Lumbocapsular pathway	2
	2) Periureteric pathway	2
	C) Stenosis of the abdominal aorta	
	1) Internal mammary pathway	
	1) Internal mammary pathway	4
	2) Pathway through meandering mesenteric artery	2
	3) Lumbar pathway	2
	4) Inferior intercostal pathway	1

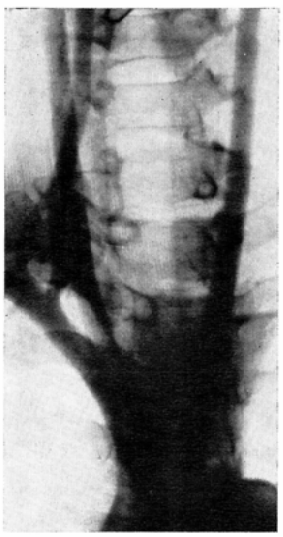


Fig. 9. Occlusion of the subclavian artery at its origin. The vertebral artery not visible on left side, while slightly dilated on right side. Arch aortogram (1.5 sec.).

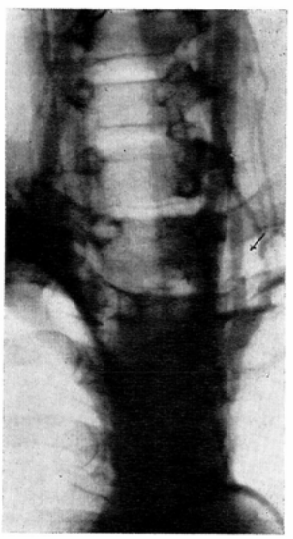


Fig. 9. b) Retrograde flow through the left vertebral artery (→) into the left svbclavian artery. (3.0 sec.)

served as the major by-pass route when the subclavian lesion was distal to the origin of the thyrocervical trunk (Fig. 3). This was seen in 24 cases (rt. 16, lt. 11, total sites 27).

(b) Reversed vertebral flow

Fig. 10. Dilated intercostal arteries in subclavian occlusion.

When occlusion of the subclavian artery at a point distal to the origin of the vertebral artery occured, a reversed vertebral flow in the ipsilateral vertebral artery was noted to supply the distal subclavian artery (Fig. 9). This was seen in 7 cases, all on left side. Four of these had symptoms of cerebral ischemia and could be called "subclavian steal syndromes"[14)38)].

(c) Aortic intercostal pathway

Well developed aortic intercostal arteries served as an effective collateral route in 13 cases of subclavian occlusions (rt. 8, lt. 11, total sites 19). (Fig. 10).

(d) Anterior intercostal pasthway

Blood flow of the occluded subclavian artery was by-passed through the anterior intercostal arteries via the internal mammary artery in 5 cases, all on right side.

(e) Inferior thyroid pathway

Anastomosis between the two thyrocervical trunks via the inferior thyroid arteries was seen in 7 cases.

(f) Pathway through the "cervical collateral network"

A well known anastomosis that links the carotid, subclavian and vertebral arteries has been referred to as the "cervical arterial collateral network"[9)], consisting of the occipital branches of the external carotid artery, the muscular branches of the vertebral artery and the thyrocervical and costocervical trunks of the subclavian artery. The direction of the blood flow may be cephalad in case of carotid occlusion and caudad in case of subclavian occlusion. This route was observed in 14 cases (rt. 5, lt. 9, total sites 14). (Fig. 3).

(g) Pathway through the "Superior thoracic collateral network"

A characteristic anastomotic network, previously not described, was observed around the aortic arch in 16 cases. This network utilized superior intercostal arteries, internal mammary artery and numerous small branches near the arch. (Fig. 11). This network, referred to in this study as "Superior thoracic collateral network", showed a high incidence in cases with extensive involvement of more than two branches of the arch, especially of the innominate and left common carotid arteries. Bolld flow via this route mainly supplied the carotid and vertebral regions and partially the subclavian region.

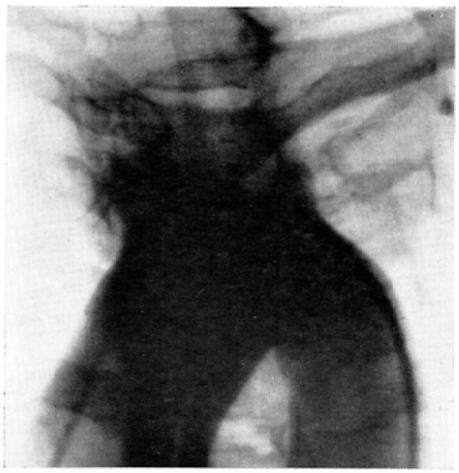


Fig. 11. a) Complete occlusion of innominate, left common carotid and left subclavian artery. Thoracic aortogram (1.0 sec.).

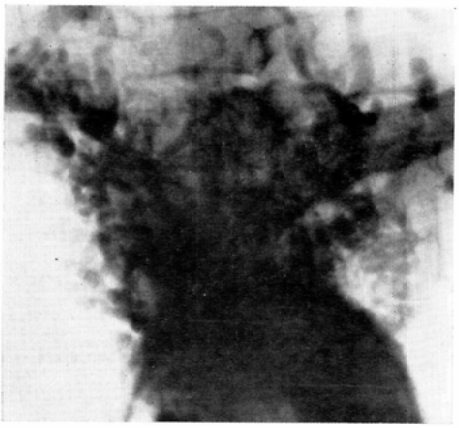


Fig. 11. b) Characteristic appearance of "Superior thoracic collateral network". (2.5 sec.).

(2) Collaterals in Celiac or Mesenteric Occlusion

(a) Pathway through the "Meandering mesenteric artery"

The meandering artery —an enlargement of a normally existing central anastomotic artery or marginal artery of the colon— was demonstrated in 11 cases. (Fig. 12). Of these, 8 were seen in occlusion of the superior mesenteric artery, 1 in occlusion of the inferior mesenteric artery and 2 in occlusion of the abdominal aorta.

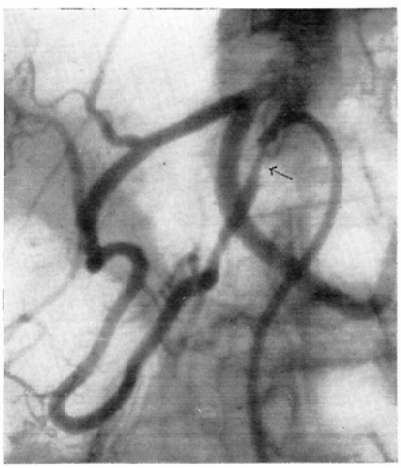


Fig. 12. a) Selective injection into the superior mesenteric artery. (0 sec.). Marked stenosis of superior mesenteric artery (→). Celiac artery is occluded and supplied via the superior mesenteric artery.

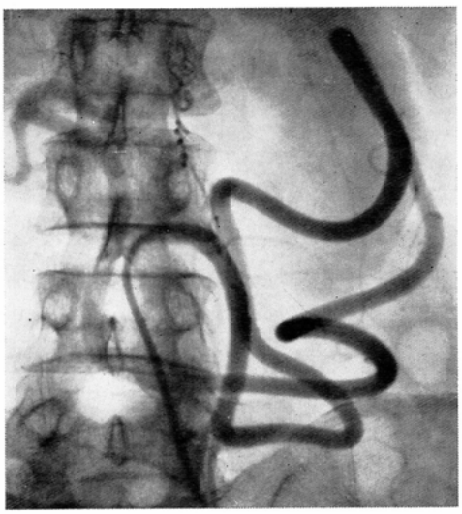


Fig. 12. b) Selective injection into the inferior mesenteric artery (0 sec.).

Fig. 12. c) (1.0 sec.). The "meandering mesenteric artery carries the blood from the inferior mesenteric artery to the superior mesenteric and then to the celiac artery.

(b) Pathway through the "Pancreatico-duodenal arcade"

In 2 cases of superior mesenteric occlusion and in 1 case of celiac artery occlusion, a significant collateral circulation via the pancreatico-duodenal arcade (the anastomosis of the superior and inferior pancreatico-duodenal arteries around the head of the pancreas) was seen.

(c) Pathway through the "Bühler's artery"

A collateral route directly connecting the celiac artery to the proximal portion of the superior mesenteric artery (Bühler's artery) was observed in 1 cases.

(3) Collaterals in Renal Artery Stenosis

In 16 cases of renal artery stenosis, a significant collateral circulation was demonstrated in only 3 cases. These were lumbocapsular pathway in 2 cases and periureteric pathway in 2 cases. One of these 3 cases demonstrated both lumbocapsular and periureteric pathways.

(4) Collaterals in Stenosis of the Abdominal Aorta

(a) Internal mammary pathway

In 4 cases of complete or incomplete occlusion of the abdominal aorta a large tortuous internal mammary artery served as a collateral route to the lower half of the body, probably coursing from the inertnal mammary to the superior epigastric to the inferior epigastric to the external iliac artery.

(b) Lumbar pathway

Dilated lumbar arteries carried the blood flow to the internal and external iliac artery in 2 cases anastomosing with the iliolumbar and deep circumflex iliac artery.

(c) Inferior intercostal pathway

In 1 case of complete occlusion of the abdominal aorta dilated lower intercostal arteries apparently served as effective collaterals and supposedly entered the external iliac artery via the deep circumflex and inferior epigastric arteries.

(d) Pathway through the meandering mesenteric artery

As stated in (2), (a) above, this pathway was seen in 2 cases.

Discussion

Nasu[28], based on his pathological observations, concludes that pulseless disease is essentially mesarteritis of the aortic arch and major arteries arising from the aorta. It is characterized by a granulomatous or diffuse productive inflammation of the media and adventitia by way of vasa vasorum with secondary juvenile arteriosclerosis. Angiographic appearances of these changes are variable. Of these, stenosis and dilatation seems to be the principal forms of abnormalities, although the latter is rare in branch lesions.

Occlusive lesions of bracheocephalic arteries are the cause of absent or diminished pulses in the neck or arm. Angiographic features of these lesions have been described by Grollman[19] as flame-shaped occlusions and smooth-localized constrictions. The regular nature of the constriction and the smooth walls within the occluded region were stressed by Wickbomb[41] as the important signs in differentiating this condition from arteriolscerosis. In our patients, however, the incidence of irregular walled constrictions is about equal to that of smooth walled lesions. It is not clear if this irregularity of the wall represents another feature of the disease or that it simply reflects a presence of marked secondary arteriosclerosis.

Attention has been called to the renal artery stenosis in connection with the high incidence of hypertension in pulseless disease. There is good evidence that approximately 50% of renovascular hypertension in Japanese are caused by aortitis of the Takayasu type[39]. The reported incidence of renal artery stenosis is 20 out of 29 by Kozuka et. al[24]., and 14 out of 36 by Tasaka[37]. As to the mechanism of hypertension in pulseless disease, Ask-Upmark[4] mentioned four possibilities: 1) Reduced elasticity of the walls of the arterial "wind kettle", 2) Reduced blood supply to the brain, 3) Elimination of the buffer nerves, 4) Interference with the renal blood supply. He finally concluded that "the main cause of the hypertension seems to be renal". In the author's series, the incidence of hypertension in 16 cases with renal artery stenosis is significantly higher than in those cases which have no stenosis. However, almost all cases of renal artery stenosis were associated with stenosis of the aorta or aortic insufficiency and these two factors seemed to correlate better with the incidnce of hypertension than renal artery stenosis. (See Fig. 6). It is reasonable to assume that the elasticity of the "wind kettle" is greatly reduced in cases of severe aortitis and extensive bracheocephalic occlusion. Most of the hypertensive cases show an increase in systolic pressure with normal or decreased diastolic pressures. This type of hypertension has been considered characteristic of the "wind-kettle type", whereas in renovascular hypertension a rise in diastolic pressure is an important feature. Hypertension in the presense of aortic insufficiency is another important finding which Usk-Upmark did not mention. It seems evident that the cause of hypertension in pulseless disease is not simply renal, but rather a complex combination of various factors discussed above. In fact there may be other factors in addition to these.

As coronary arteriography was not performed in this study, coronary lesions were not definitely proven except for 1 case in which marked aneurysmal dilatation of the left coranary artery was clearly demonstrated on thoracic aortograms. Association of myocardial infarction in cases of pulseless disease has been reported,[5,11] but angiographic demonstration of the coronary lesions has not been reported in the literature. Coronary arteriography may be indicated in cases with positive ECG findings or other signs of ischemic heart disease.

Stenosis of the aorta, in most of the cases, takes the form of "elongate coarctation"[26]. It is chara-

cteristically long stenotic segment with smooth or irregular walls and has a predilection for the lower portion of the descending aorta. Isthmus stenosis, such as was seen in one case in the present study, might be a rare exception.

The incidence of aneurysm of the aorta in pulseless disease has been reported as 8 to 16 per cent[30,36]. In this study it was 4 per cent. The finding of an aneurysm in younger patients, particularly in Japan, should bring to mind the possibility of pulseless disease or aortitis. Thickening of the wall of the aorta has been mentioned as an interesting feature of this disease[15,36]. This is most clearly demonstrated in the left wall of the descending aorta on thoracic aortograms and hardly seen in the other portion of the aorta.

The occurence of aortic insufficiency in pulseless disease is a grave prognostic sign since it is often associated with hypertension and cardiac failure. The first report of such a case was made by Jervel[22] in 1954. Aortic regurgitation seems to be due to marked dilatation of the ascending aorta resulting in commissural separation, and is probably not due to direct involvement of the aortic valve with arteritis. This view is supported in one autopsied case in the present series. Several reports by different authors offer a similar view.[11,30,42]

Frøvig and Løken[17](1951)demonstrated intercostal collateral routes by post-mortem angiography using barium paste. Recent developement of improved angiographic technique has made it possible to analyze the pattern of collateral circulation more completely. However, no comprehensive description of the various collateral routes in pulseless disease is found in the literature. In proximal subclavian occlusion due to arteriosclerosis, reversed vertebral flow (Subclavian steal) is the most frequent and the most effective collateral route. In pulseless disease, however, the incidence of this route is not high. Seven out of 32 cases with proximal subclavian lesions in the present series showed reversed flow in the ipsilateral vertebral artery. This incidence is low compared with that reported by Newton[29](12 out of 15) and Heyman[20] (7 out of 14), both in cases of arteriosclerotic occlusions. Lang[25] by his quantatative method using radioisotopes, showed an interesting fact which might explain the low incidence of the subclavian steal in pulseless disease. According to his observation, in young patients with surgical ligation of the subclavian artery following a Blalock-Taussig procedure, the predominant collateral supply is initially via the vertebral system. Serial follow-up isotope studies, however, demonstrate a progressive decrease of retrograde flow via the vertebral artery and a progressive increase of antegrade collaterals via the internal mammary and or intercostal arteries. Twenty four moths after the ligation, the predominant flow is via the antegrade collaterals. He also pointed out that this dynamic inversion between the vertebral and the other antegrade collaterals was not observed in older patients with arteriosclerotic occlusion of the subclavian artery. As to the terminology some[10] insist that distinction should be made between the true "subclavian steal syndrome" in which reversed vertebral flow is associated with cerebral ischemic symptoms and "subclavian steal pattern" in which no cerebral symptoms are seen. Others[1,29], however, seems to make no such strict distinction between the two. Actually in patients with cerebral ischemic symptoms, it is not always possible to determine whether these symptoms are the result of a subclavian steal or are the result of occlusion of carotid arteries. In cases of distal subclavian occlusion, the predominant collateral route is via the branches of thyrocervical trunk. Intercostal arteries theoretically posess the ability to act both as an antegrade collateral to the subclavian lesion and as a retrograde collateral to the aortic lesion. Actually, however, no case with a retrograde intercostal route was observed in the author's cases.

Sen[33] reported similar findings in 14 cases of stenosis of the thoracic aorta due to aortitis. It may be that in cases with stenosis of the sorta the inflammatory process is so extensive as to obliterate the origin of the intercostal arteries. Another reason for this may be that no case had complete occlusion of the thoracic aorta necessitating large amounts of collateral flow through the intercostal arteries. In connection with this, a low incidence of rib notching in chest film of patients with pulseless disease has been pointed out.[8,24] In the present series, rib notching was not remarkable even in the cases with extensive enlargement of intercostal arteries, being observed only in 4 cases. A characteristic form of collateral vessels, previously not described, is referred to in this study as "superior thoracic collateral network". This is a complex anastomosis between the superior intercostal artery, internal mammary artery, and the numerous unnamed small arteries around the aortic arch and frequently observed in cases with extensive involvement of the aortic arch branches. Among the collaterals in the abdominal region, the presence of a meandering mesenteric artery[27] is the most interesting. This is an enlargement of normally existing channel between the superior and inferior mesenteric artery and is able to serve both in occlusion of the abdominal aorta and in occulsion of its visceral branches. Tasaka[38] reported that 13 cases in a total of 270 abdominal aortography had this channel, and of these, 9 were cases of pulseless disease. Occlusive changes of the celiac artery or mesenteric arteries are often suggested by the meandering artery and careful observation of the direction of the flow on serial roentgenograms is of prime importance.

As to the technical aspect of the examinations, percutaneous thoracic and abdominal aortography using automatic pressure injector and serial exposure system is the most reliable procedure. Abdominal aortography should never be ommited since more than 40 per cent of the cases show some angiographic abnormalities in the abdominal region. In advancing the catheter under fluoroscopic control, the possibility of stenosis or aneurysm of the aorta should always be born in mind in order not to damage the wall of the aorta. Selective injection of the branches of the aorta affords more clear and detailed informations concerning pathological condition, however, care should be taken as this is not without risk in the presence of severe arteritis.

Summary

Thoracic and abdominal aortography were performed in 50 cases of pulseless disease during a period of 3 years and 4 months. In 17 of these, additional selective arteriography of the major branches of the aorta was also performed. All the angiographic abnormalities were presented and a classification of the cases according to the aortic abnormality was given. Special referrence was paid to collateral circulation in this disease and various patterns and frequency of the collateral vessels were presented including a new form of collateral network, previously not described. Pertinent literature was reviewed and discussed.

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