



FIG. 6.—Diagram of the operation of portal-systemic exchange.

The immediate cause of Banti's syndrome is generally thought to be a damming back of the blood in the portal venous tree.

The portal-vein obstruction may be extrahepatic (12 cases) or intrahepatic as a result of cirrhosis (56 cases).

A cirrhotic liver is not necessarily associated with Banti's syndrome, but the conditions overlap to such an extent that it is advisable, in the present state of our knowledge, to consider them as one group.

The indications for operation are: (a) gastro-oesophageal haemorrhage; (b) hypersplenism; (c) ascites, after medical treatment has failed; and (d) associated conditions, such as biliary obstruction.

When the liver is normal or only mildly cirrhotic the operation of splenectomy (to remove the overactive spleen) and spleno-renal anastomosis (to reduce the portal venous pressure) gives excellent results, provided that (a) the splenic vein is patent, and (b) the veins are of sufficient size.

Portal venography is of great value in demonstrating the exact state of the portal venous tree.

In the case of one patient with extrahepatic obstruction who had no available splenic vein, resection of the lower 4 in. (10 cm.) of the oesophagus and proximal two-thirds of the stomach has proved successful.

Porta-caval anastomosis provides an alternative and most efficient method of reducing portal venous pressure, and is clearly indicated for gastro-oesophageal haemorrhages in the absence of hypersplenism.

Portal-systemic venous anastomosis should be done early in the course of the disease because it is hazardous in cases of advanced cirrhosis. Careful clinical and biochemical assessment is necessary to select cases suitable for this operation. Those that survive are much improved.

All venous anastomoses should be done by the suture method, with intima-to-intima approximation.

For the advanced cirrhotic patients who are considered unsuitable for a shunt the operation of common hepatic and splenic arterial ligation offers hope.

The case histories of a complete consecutive series of 68 patients are set out in tables, in which an attempt has been made to relate the underlying cause, the symptoms, and the duration of the disease to the treatment and the result.

Cirrhosis in the terminal stages has not responded to any form of treatment. A method of replacing the portal blood flow to the liver is briefly described, but this operation failed in four cases.

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SYMMETRICAL ARTERIAL OCCLUSION OF UPPER EXTREMITIES, HEAD, AND NECK: A RARE SYNDROME

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We have recently had under our observation two almost identical cases of a very unusual circulatory anomaly. The main effect of the anomaly is to cause profound restriction, in symmetrical fashion, of the blood supply to the upper extremities, head, and neck, with the production of certain symptoms. Our interest in these patients was aroused by noting, first, that the radial pulse was absent at both wrists, although the radial arteries themselves could be felt in the normal position; and, secondly, that pulsation was virtually absent in all the arteries of the upper limbs, head and neck.

Apart from embolism, trauma, and anatomical variations in the course of the radial artery, unilateral absence of the arterial pulse in an upper extremity is a rare phenomenon, and has been found chiefly in syphilitic aneurysm of the aorta and in dissecting aneurysm. Bilateral absence of the pulse is rarer still, and we have been able to collect only 16 cases from the literature. In examining the records of these we were at once struck with their close resemblance to our own patients. Thus in the great majority the arterial pulse was lacking not only at the wrist but in the upper extremities, and in the head and neck; indeed, the symptoms and clinical findings in the series of cases as a whole formed a remarkably constant pattern. The purpose of this paper is to discuss this syndrome, as it appears to be little known. Our own cases are first described.

Case 1

A colliery lamp-man aged 42 was admitted on June 6, 1948. He had previously been in excellent health; but in January, 1948, he began to notice that his vision became blurred for about ten minutes at a time. He also experienced momentary faintness on standing and walking, usually accompanied by blurred vision, several times each day, and relieved at once by sitting down. He lost consciousness for a few moments on one occasion when being tilted backwards in a barber's chair. The visual disturbance and faintness recurred frequently, often several times a day, particularly on bending his head backwards or when wearing a tight collar, but there were no other symptoms, and he was an active man, although his right leg had been amputated below the knee at the age of 17 following an injury to his foot. His only previous illness had been an attack of rheumatism at the age of 12. There was no known Jewish ancestry, and he was a light smoker. There was no family history of hypertension.

On examination he was thin, pale, and of rather poor general physique; weight 7 st. (44.5 kg.). The pulse could not be felt at either wrist and was barely discernible in the left brachial, axillary, and common carotid arteries. It was absent from these vessels on the right side, and from both subclavian arteries. The arteries themselves in the upper limbs could easily be palpated in their normal positions. The blood pressure could not be registered with a Baumanometer in either arm. Oscillometric readings: maximum pulsation right arm, 10 mm. at a pressure of 90 mm.; left arm, 15 mm. at 120 mm. A small aberrant vessel could be felt in the left posterior triangle of the neck, and pulsating arteries were present over the back. Pulsation in the abdominal aorta and in the arteries of the lower limbs seemed normal. The blood pressure in the right leg averaged 200/130; in the left leg, 240/140. Oscillometric readings: maximum pulsation right leg, 25 mm. at 150 mm.; left leg, 30 mm. at 140 mm. Examination of the heart and an E.C.G. were normal. The circulation in the skin was good. Ophthalmoscopic examination revealed no abnormality in the lenses, but a visible circulation was present in the main branches of the retinal arteries, the blood passing along the arteries in an even slow stream without pulsation. This phenomenon was observed particularly at such times as the patient was feeling faint, and was seldom seen in two vessels simultaneously. The retinal veins were dilated.

X-ray films of the heart and lungs were normal, and rib notching could not be seen. An angiogram showed normal filling of the heart, pulmonary arteries, and aortic arch. The last named seemed normal in shape, size, and position, and there was no sign of stricture. At least two of the great arteries arising from the arch could be visualized at their origins, and seemed of normal calibre, but they could not be traced very far. X-ray films of the skull and extremities were normal. The urine contained no abnormality, the blood urea was 30 mg. per 100 ml., and the urea-clearance test gave figures of 46% and 37%. An intravenous pyelogram was normal. The Wassermann reaction was negative, and the blood count was normal. A biopsy of the left brachial artery at three levels showed it to be fully patent and of average size. Sections of it were normal. The patient was discharged from hospital on July 23, 1948, and returned to work six months later.



FIG. 1.

FIG. 2.

FIG. 1.—Case 1. Showing cataract formation and wide dilatation of the pupil of the affected eye. FIG. 2.—Case 2. Showing cataract formation and wide dilatation of the pupils. The newly formed circular vessel at the base of each iris is just visible.

When next seen in May, 1951, he stated that the attacks of blurred vision had continued, and that during the preceding few months vision in the right eye had gradually failed. The right eye was now practically blind, a complete cataract having formed with, also, atrophy of the iris (Fig. 1). At the base of each iris the beginning of superficial vascularization could be seen. The right pupil was widely dilated. In the left eye there were incipient lens opacities, and the fundus showed some minute haemorrhages and small aneurysmal dilatations here and there in the terminal arterial branches. Light pressure on the ball of the left eye caused collapse and blanching of all the retinal vessels. The patient's condition was otherwise as before, and further x-ray films showed that no cardiac hypertrophy had developed.

Case 2

A married woman aged 37 was admitted on February 22, 1949, under the care of Mr. Hardman, complaining of failing eyesight. Her childhood and adolescence had been uneventful except for very occasional fainting attacks, to which she paid little attention. She began training as a nurse at the age of 19 and used to feel her own pulse occasionally "for practice." She married in 1935, and in 1936 was delivered of a living child five weeks before term. Throughout this pregnancy she suffered from frequent "faint turns" each day but did not fall. These symptoms disappeared after delivery, but returned in 1937, when she was again pregnant. During this pregnancy, which resulted in the birth of another premature child surviving two days, the brief syncopal attacks occurred usually once a week, and continued with about the same frequency after delivery.

In 1944 the uterus was removed because of abdominal pain. Immediately after this operation she began to suffer from severe generalized headaches, and on the thirteenth day a sudden right hemiplegia involving the face occurred. Simultaneously she became almost completely blind in both eyes, but within a week her vision recovered. The headaches continued, but the hemiplegia slowly improved, and within a few months all weakness had disappeared. In 1947 the vision in her right eye suddenly began to fail, at first for a few minutes at a time but later for increasingly longer periods. This phenomenon occurred almost daily, and in November, 1948, she lost the vision in both eyes for several hours. Thereafter, her eyesight deteriorated, with exacerbations of complete blindness, whilst the fainting attacks became much more severe. She began falling to the ground many times each day, though consciousness was only momentarily lost. Her doctor was unable to feel her pulse or to record her blood pressure. She was a light smoker and had no known Jewish ancestry.

Her general condition and colour were good and her intelligence average; weight 7 st. 12 lb. (49.9 kg.). Pulsation could not be felt in either carotid, subclavian, brachial, or radial arteries, but these arteries could be felt in their usual positions. The blood pressure could not be obtained in the arms with a Baumanometer. Oscillometry: maximum reading 5 mm. and 2.5 mm. in the right and left upper arms respectively at a pressure of 70 mm. Pulsation in the abdominal aorta and femoral arteries was strong and the blood pressure in the lower limbs was 150/90. Oscillometry: maximum reading 10 mm. and 15 mm. in the right and left thighs respectively at a pressure of 150 mm. Many collateral pulsating vessels could be felt over the back of the chest, a bruit being heard over several. A loud continuous murmur, most pronounced in systole, was present over a localized area just above the inner end of the left clavicle. Peripheral skin circulation was normal. The heart, lungs, and abdomen were normal clinically.

Her vision was poor, and she was unable to read big news print. Both fundi showed many exudates and haemorrhages in all layers. There was considerable venous engorgement, and some segmentation of the

circulating blood in the veins was at times visible, the blood appearing to move in clumps. The arterial circulation could not be seen. At the base of each iris, running round in approximately the position of the contraction groove, was a newly formed circular vessel. The pupils were dilated, and did not react to light or accommodation, but the lenses appeared healthy. Examination of the rest of the nervous system was negative. Electrocardiograms and x-ray films of the heart and lungs were normal except for the presence of small bilateral cervical ribs. The cerebrospinal fluid was normal. The Wassermann reaction was negative in the blood and cerebrospinal fluid. The urine contained no abnormality, and dilution and concentration tests were normal. A urea-clearance test gave figures of 53% and 55%, and an intravenous pyelogram was normal. A blood count showed a mild hypochromic anaemia.

On March 19 Mr. Hardman performed a cerebral angiogram; 50% diodone was injected into the left common carotid artery between the heads of the sterno-mastoid muscle. Great difficulty was experienced in inserting the needle, and subsequent x-ray films suggested that the vessel possessed a small lumen. The external carotid did not fill, but the vessels of the brain were well visualized. After this procedure the patient became aphasic, but slowly recovered. Portions of the right radial and left temporal artery were excised. These vessels appeared to be smaller than the average, but their lumina were patent and no pathological changes were found in their walls. She was discharged on March 27.

She was readmitted, under our care, on September 24, 1949. She was now blind, and bilateral cataracts had formed, with marked atrophy of each iris and ciliary body (Fig. 2). The peripheral iritic vessels already mentioned were now replaced by a pigmented fibrous ring around the base of the iris in each eye. There was some degenerative keratitis in the left eye, with superficial vascularization. Her headaches, especially above the left eye, had continued to be severe, but the syncopal attacks had decreased in frequency and were now occurring only once every few weeks. She was sent home on October 19, but remained under periodic observation, and in September, 1950, had shooting pains in both legs which were much worse on walking. Her lower limbs had begun to feel cold, and the arterial pulse below the thigh was now difficult to feel, whilst it was not possible accurately to record the blood pressure in the lower limbs with a sphygmomanometer. Oscillometry showed a fall in the maximum reading in each thigh to 5 mm. at a pressure of 150 mm. No trophic changes were apparent.

Aetiology

The physiological result of the lesion in both our cases is to impair the circulation of the cerebrum to a degree sufficient to cause syncopal attacks and visual disturbance, and it seems clear that the lesion, whatever its nature, must be either in the arch of the aorta, involving the orifices of the great arteries arising therefrom, and/or in these arteries themselves. The cases of bilateral absence of the pulse which we have collected from the literature, together with our own cases, are summarized in the Table. In the first group (Cases 1-9) there was strongly presumptive or certain evidence of syphilis; in the second group (Cases 10-18) this disease can be excluded with reasonable certainty, with the possible exception of Cases 13 and 14.

Cases of Syphilitic Origin

An aortic aneurysm was found in four of the five fatal cases, but was apparently the direct cause of death in only one instance (Türk, 1901). Broadbent (1875), who seems to have recorded the first case of bilateral absence of the pulse, found the aorta to be of healthy appearance at necropsy, although the patient had had syphilis. He found narrowing, rigidity, and crowding together of the orifices of the great vessels arising from the arch, and concluded that atheroma played a part in causing the obstruction. Shikhare (1921)

reported that the aneurysm in his case contained an ante-mortem clot which sent finger-like projections into the great arteries, causing obstruction. In the other cases, however, the arterial orifices were involved in the syphilitic mesoaortitis, while syphilitic changes were also in some instances demonstrated in the proximal portions of the great arteries for a variable distance. Sato (1938) demonstrated thickening, narrowing, and absent pulsation in these arteries by dissection during life. Töppich (1921), in an extensive review, drew attention to a collar-like thickening around the origins of the aortic arch branches in certain cases of syphilitic aortitis, and showed that the actual obstruction is caused by growth of the aortic intima across the mouths of the arteries.

Cases of Non-syphilitic Origin

It is in this group of cases, which includes our own patients, that uncertainty arises concerning the cause of the arterial obstruction. Only scanty necropsy material is available, and further study is obviously necessary.

Atheroma

Bittorf (1947) and Marinesco and Kreindler (1936) thought atheroma to be the cause of the obstruction in their cases. At necropsy the latter authors found extensive atheroma with calcification of the aorta and complete obliteration of the orifices of the great arteries arising from its arch. The innominate was obstructed by organized thrombus, which also filled both subclavian arteries as far as the origins of the internal mammary arteries. Histologically, the left common carotid showed atheromatous changes, the lesion being typical of "endarteritis obliterans" without evidence of any specific inflammatory reaction.

It seems possible that atheroma may sometimes be responsible for the obstruction, for atheroma of the aorta is often especially marked in relation to the origins of the arteries arising from it, and obliterating arteriosclerosis may occur in the upper limbs, although it is uncommon. Thus Allen, Barker, and Hines (1946) state that, rarely, atheromatous formation at the origin of the subclavian artery may lead to extensive thrombosis in this vessel. Moreover, as atheroma often complicates luetic mesoaortitis, it may sometimes play a part in the production of the obstruction when the basic lesion is syphilitic, as was suggested by Cohen and Davie (1933). We do not think it at all probable that atheroma is the cause of the obstruction in our own patients, in view of their ages and of the normal histological findings in the excised portions of the peripheral arteries.

Arteritis

In two cases there was evidence of arteritis. Harbitz (1926) found total occlusion of the subclavian arteries and of the common carotids as far as their bifurcation, while the innominate was almost completely blocked. The affected vessels were filled with organized blood clot, and marked inflammatory changes were present in the common carotids, consisting of extreme infiltration of the media with lymphocytes along the vasa vasorum. Giant cells were not seen. Inflammatory changes were less marked in the internal carotids, which showed considerable intimal thickening. The aorta was inelastic, with patches of calcification, while its arch, and to a less extent the descending aorta, showed extensive round-celled infiltration of the adventitia, less marked in the media. The abdominal aorta and femoral arteries were normal. No organisms could be demonstrated, but a possible infective origin was postulated.

Froving (1946) investigated his patient, a woman aged 21, in great detail. Attempts at angiography through the left common carotid failed, this vessel on exposure being found hard, stiff, and pulseless. Injection of the left subclavian artery proximal to its vertebral branch was also unsuccessful, though good filling of the arm arteries was obtained through the middle portion of the left brachial artery. This vessel was also pulseless and seemed small. The right common carotid resembled the left macroscopically. Sections of it

Summary of Known Cases with Bilateral Absence of Pulse

Case No.	Author	Sex and Age	Chief Symptoms	Duration in Years	W.R.	B.P. Arms	B.P. Legs	Clinical Features
<i>Cases of Syphilitic Origin</i>								
1	Broadbent (1875) ..	M 50	Referable to hepatic cirrhosis	?	?	?	?	History of syphilis. Absence of radial pulses for 30 years. Vigorous pulse in femoral arteries. Died
2	Türk (1901) ..	M 44	Transient faintness; blurred vision; angina of effort; weakness of arms	1	?	Nil	?	History of syphilis. Feeble pulse in right carotid and right subclavian. Pulse absent in other arteries of arms, head, and neck. Strong pulse in abdominal aorta and femorals. Collateral circulation. Pronounced arteriosclerosis. Rupture of aneurysm into oesophagus. Died
3	Shikhare (1921) ..	M Middle-aged	Frequent giddiness and syncope	?	?	?	?	Absent pulse in all arteries of upper part of body. Normal pulse in abdominal aorta and legs. Aneurysm of aortic arch. Died
4	Crawford (1921) ..	M 52	Dizziness; severe headaches; dyspnoea; cyanosis	1½	Pos.	Nil	125	Absent pulse in radial, brachial, and carotid arteries; forcible in abdominal aorta; normal in femorals. Clinical and radiological evidence of aneurysm of arch and descending thoracic aorta
5	Kampmeier and Neumann (1930)	M 35	Giddiness and frequent syncope; recurrent visual failure; pains in chest	6	Pos.	Nil	178/110	Absent pulse in radial, brachial, and carotids. Unilateral absence of radial pulse for 4 years previously. Good pulse in abdominal aorta and legs. Collateral circulation. Clinical and radiological evidence of aneurysm of thoracic aorta
6	Cohen and Davie (1933)	M 60	Sudden blindness left eye; recurrent syncope, and headaches	12	Pos.	40-50 (oscillometer)	210	Absence of radial and carotid pulses. Collateral circulation. Clinical and radiological evidence of aneurysm of thoracic aorta. Cataract left eye. Delirium and hemiplegia. Died
7	Sato (1938) ..	F 25	Giddiness; frequent fits; headaches; gradual deterioration of vision; fatigue of upper extremities	1	Pos.	Nil	125/85	Absent pulse upper extremities, head, and neck. Normal pulse abdominal aorta and legs. Normal heart x-ray film. Atrophy of brain revealed by air encephalogram. Bilateral cataract, atrophy of iris, arteriovenous anastomoses of retinal vessels
8	Lampen and Wadulla (1950)	F 52	Giddiness and syncope; deterioration of vision; dyspnoea on exertion	1	Pos.	Nil	270/90	Pulse absent in arms and left carotid. Weak in right carotid. Strong pulse in abdominal aorta and legs. Radiological evidence of diffuse dilatation of aorta. Slow circulation in retinal vessels, exudates at left macula, and lowered retinal pressure. Hands white and cold. Cerebral softening. Died
9	Lampen and Wadulla (1950)	F 52	Repeated syncope; recurrent dimness of vision; dyspnoea and swelling of feet	2	Pos.	70/40	290/100	Pulse absent in arms and right carotid. Feeble in left carotid. X-ray films showed diffuse dilatation of aortic arch and fusiform enlargement of descending aorta. Hands white and cold
<i>Cases of Non-syphilitic Origin</i>								
10	Harbitz (1926) ..	F 37	Recurrent dimness of vision; blindness right eye; jerking of limbs; fatigue of limbs	1½	Neg.	?	?	Bilateral absence of radial pulse. Slow retinal circulation. Cataract of right eye. Atrophy of iris. Pigmentation of face with atrophy of right side of face. Perforation of nasal septum. Right hemiplegia and coma. Died
11	Marinesco and Kreindler (1936)	F 38	Recurrent dimness of vision; progressive loss of vision left eye; headaches, epileptiform attacks, left hemiparesis	9	"	Nil	140/90	Absent pulsation arms, neck, and face. Normal heart x-ray film. Atrophy of bones of face, with pigmentation. Perforation of nasal septum. Progressive cachexia and cerebral softening. Died
12	Giffin (1939) ..	F 19	Severe headaches; attacks of blurred vision	Several	"	"	190/110	Pulse absent in arms and left carotid, diminished in right carotid. Increased pulsation in abdominal aorta and legs. No radiological evidence of aneurysm, but pulmonary conus enlarged and aortic knuckle absent. Fragmentary circulation in retinal veins, with lowered retinal pressure. Fatigue of jaws and upper limbs
13	Oota (1940) ..	F 25	?	?	?	?	?	Absent pulsation in carotid, brachial, and radial arteries. "Peripapillary anastomoses in eyes." Few details given
14	Takahasi (1940) ..	F 28	Frequent syncope, dizziness, and headaches; delirium	7	?	?	?	Absent pulsation in carotid and radial arteries. Cataract in left eye. Few clinical details
15	Froviq (1946) ..	F 21	Syncopal attacks, recurrent blurring of vision, blindness of right eye; right followed by left hemiplegia	1	Neg.	Nil	230/150	Pulse absent in arms, neck, and face. Normal heart x-ray film. Air encephalogram showed atrophy of left cerebral hemisphere. "Granular" circulation in retinal vessels, with retinal haemorrhages and lowered retinal pressure. Right cataract; atrophy of iris
16	Bittorf (1947) ..	M 56	Recurrent syncope; transient diminution of vision right eye; attack of blindness lasting 10 minutes	9	"	60/40 Later nil	?	Carotid and subclavian pulses barely felt, brachial and radial pulses absent. Femoral pulses strong. Arteriosclerosis. Collateral circulation. No radiological findings given. No evidence of aneurysm
17	Skipper and Flint ..	M 42	Recurrent faintness and blurred vision; blindness left eye	3½	"	Nil	240/140	Pulse absent in both radials, right brachial, and right common carotid, barely perceptible in left brachial and left common carotid. Normal pulse in abdominal aorta and legs. No radiological abnormality in chest. Collateral circulation. Visible circulation in retinal arteries. Right-sided cataract, incipient opacities left lens. Atrophy of each iris
18	Skipper and Flint ..	F 37	Severe headaches, hemiplegia, recurrent syncope, and blurring of vision progressing to blindness; intermittent claudication in legs	15	"	"	150/90 Later nil	Absent pulse in arms, neck, and face. Systolic bruit above left clavicle. Normal pulse in abdominal aorta and legs; pulse in legs later difficult to feel. X-ray film of chest normal. Collateral circulation. Fragmentation of retinal circulation. Bilateral retinopathy, cataracts, and atrophy of each iris

showed great swelling and cell proliferation of the intima and fibroblastic proliferation in the media with the formation of a few giant cells. There were small clusters of round cells, particularly in the boundary zone between the adventitia and media.

The cases considered in this paper bear little clinical resemblance to the well-recognized varieties of arteritis. Temporal arteritis runs a different course, and only in its visual phenomena is there any similarity; while according to Harrison (1948) only 2 of the 75 recorded cases have occurred in young subjects. One of these, that of a woman aged 23, was published by Gilmour (1941) as a case of giant-celled arteritis. The temporal arteries were not mentioned and were presumably not involved, but Gilmour considered the pathological changes to be similar to, or identical with, those of temporal arteritis. There was obstruction of the right subclavian and right common carotid, but we have found no other instance of this disease in which occlusion of a great artery has occurred. Thromboangiitis obliterans is rare in women, and, although visual and cerebral symptoms may occur when it involves the arteries of the brain, there is no authentic instance, so far as we are aware, of occlusion of the main aortic arch branches in this condition. The Table shows the curiously high incidence of females below the age of 40 in the non-syphilitic group,* and this alone would seem to exclude this disease as a cause of the obstruction. Periarteritis nodosa, being a disease which chiefly affects the visceral arteries, with frequent renal involvement, need hardly be considered in the possible aetiology.

Congenital Anomaly

This was the hypothesis advanced by Giffin (1939). A patent ductus arteriosus was probably present in his case, but we do not think there was any good evidence of the existence of any of the other complex anatomical defects postulated by him, and it is difficult to see how the known congenital anomalies of the arch or its branches could account for this symmetrical type of arterial occlusion. Possibly atheroma, complicating congenitally small arterial orifices, or orifices unduly crowded together, might produce it.

To summarize, it would seem that atheroma may sometimes be responsible, but we are inclined to the belief that a form of arteritis, at present unidentifiable, which for some reason affects the aortic arch branches symmetrically, is more often the cause.

Symptomatology

Reference to the Table reveals a remarkably uniform symptomatology. As would be expected, angina and other cardiac symptoms were met with in the first group, but the presenting symptoms in both groups were almost invariably cerebral and visual. Often the symptoms were spread out over a number of years.

Cerebral Symptoms.—Faintness, giddiness, or loss of consciousness was noted in 14 cases, in all but two of which actual syncope occurred. Usually the attacks were of short duration, were worse in the upright posture, and were frequently repeated. Headache was also a prominent feature, and tended to be particularly severe when occurring with major cerebral episodes such as hemiplegia. There was evidence of gross cerebral damage, such as hemiplegia, convulsions, or cerebral softening, in seven instances.

Visual Phenomena.—These are of particular interest and are mentioned in 15 cases. One very characteristic symptom is a transient recurrent "blurring" of the vision (nine cases), occasionally without, but usually with, a progressive diminution in visual acuity. The curious slowing of the retinal circulation seen in both our cases is described in four others,

*Sato (1938) mentions five cases of absence of the radial pulse, observed in Japan, four of which were females between the ages of 15 and 25. There was progressive deterioration of vision with, it is alleged, arteriovenous anastomoses in the retinal vessels and cataract formation. As few details of his cases are given they have not been included in our series.

in three of which a lowered retinal pressure—that is, the pressure required to obliterate the retinal arteries—was present. Both Sato (1938) and Oota (1940) found "arteriovenous anastomoses" in the retinal vessels, the former also describing small haemorrhages and aneurysmal dilatations. The peripheral retinal vessels were transformed into "white strands," and there was atrophy of the choroid and retina. The extensive retinopathy in our second case has apparently not been observed in any of the others. Cataracts were found in seven cases, in two of which it was bilateral. It seems that the cataract usually forms within 12 to 18 months of the onset of the first symptom, although in our second patient the bilateral cataracts did not appear until the thirteenth year of her illness. The actual development of the cataract may be rapid, as in Frovig's case, in which it became severe in two or three days. Cohen and Davie's patient had complained of sudden loss of vision in the left eye. This was apparently due to a haemorrhage, which was followed by cataract. In five of the cases with cataract there was atrophy of the iris.

Other Symptoms.—Loss of power and ready fatigability of the upper limbs is mentioned five times, and of the jaws once. No trophic changes have been noted in the limbs, but coldness of the upper extremities occurred in both the patients described by Lampen and Wadulla (1950). Our second case is apparently the only instance of impaired blood flow to the lower limbs, and in this patient it must be assumed that pathological changes in the lower limb arteries have occurred. Harbitz (1926) described right facial hemiatrophy and Marinesco and Kreindler (1936) atrophy of the bones of the face. A curious finding in both these cases was pigmentation of the face and perforation of the nasal septum.

Abnormalities in the Circulation, and Their Relationship to Symptoms

The location and extent of the obstruction in the aortic arch branches, when verified, have already been described, and it is clear that it is the symmetry of the occlusion which is responsible for the symptoms and clinical findings. We did not feel justified, in our own patients, in making more attempts to confirm the site of obstruction by further investigation. In our first case the angiogram showed that two of the great vessels were free at their immediate origins; in our second case the obstruction in the left common carotid evidently lay between the level of the contrast injection—that is, between the two heads of the sterno-mastoid—and the origin of this vessel from the aorta. The production of the various symptoms is dependent upon the diminished blood flow through the aortic arch branches, and the slow progression of the disease suggests a gradual occlusion, though it is apparent that sudden vascular block from thrombosis may occur, for example, in the middle cerebral artery.

In all probability, the sudden hemiplegia and visual failure in our second case was due to thrombosis of the internal carotid artery, and Frovig assumed that the hemiplegia in his case was also due to this cause. The vertebral arteries must play an important part in the maintenance of the cerebral circulation. Thus Takahasi (1940) and Frovig both achieved filling of the cerebral vessels with angiograms carried out by injection of one of the vertebral arteries, while Marinesco and Kreindler filled the cerebral vessels, after death, with a mixture of gelatin and lead oxide injected into the same vessel. Normally the vertebral arteries are much less important than the internal carotids in the blood supply to the brain. If, as may happen in this disease, they become occluded, enough cerebral circulation cannot be maintained to support life. In Lampen and Wadulla's fatal case there was thrombosis of the basilar artery with cerebral softening, while Harbitz demonstrated almost complete occlusion of the vertebral arteries in addition to thrombosis of the left cerebral artery. It is evident that the visual phenomena are due to slowing of the intraocular circulation, and the formation of cataracts and other phenomena can be explained on this basis.

The circulation of the upper limbs is maintained through collateral channels, which are presumably very similar to those formed in coarctation of the aorta. Thus pulsating arteries were found over the back in four cases in addition to our own patients. The flow of blood in these anastomotic vessels must, however, be from below upwards, as was noted by both Türk and Bittorf.

The absent pulse in the upper extremities, together with the vigorous pulse and the apparently high blood pressure in the lower limbs which many of these cases exhibit, has led to the application of the unsuitable term "reversed coarctation" to the anomaly by Giffin and by Lampen and Wadulla. A systolic reading of over 200 mm. Hg was obtained in the lower limbs in five cases. With such scanty material, however, it is difficult to be sure how far these pressures exceed the normal. Wendkos and Rossman (1943) carried out a study of the blood-pressure readings in the legs of 500 normal soldiers, excluding all subjects in whom the blood pressure in the arms exceeded 140/90. While the average figure was 154/91, in 25% the systolic pressure ranged from 170 to 195 mm. Hg and in 2% from 200 to 230. They stated that there was a direct relationship between the circumference of the thigh and the height of the blood pressure, and indicated the importance of an increase in the muscle mass in raising the pressure. Nevertheless, the readings given by Lampen and Wadulla are excessive, though the high pulse pressures in these patients were not satisfactorily explained. A reading of 190/110 in Giffin's case certainly seems high for a female aged 19 who is said to have been physically underdeveloped, and a pressure of 240/140 in our first patient, who weighed only 7 st. (44.5 kg.) would also appear to be abnormal. As is to be expected, cardiac hypertrophy was noted in several of the syphilitic cases. It is not mentioned in any of those of non-syphilitic origin, though the weight of the heart in Harbitz's patient was 470 g. No discussion on the possible cause of the raised arterial tension in the lower limbs in these patients would be profitable in the present state of our knowledge of the cause of hypertension in general.

Summary

Two rare cases observed by us in which there was virtually complete absence of the arterial pulse in the upper extremities, head, and neck, owing to arterial occlusion, are described. The cause of the obstruction in these cases was not determined.

Two groups of similar cases collected from the literature are discussed. In the first, occlusion is due to syphilitic aortitis either with or without an aneurysm of the arch. In the second group the aetiology is less clear. Atheroma may be the cause in certain cases, but evidence is produced which suggests that a form of arteritis is more often responsible. The high proportion of women below the age of 40 in the non-syphilitic group renders it unlikely that thromboangiitis obliterans is the cause.

The remarkably constant symptomatology common to both groups is described, the chief symptoms being cerebral and visual. Amongst the former, repeated faintness and syncope are common, while gross cerebral damage as evidenced by convulsions, hemiplegia, or death from cerebral softening may occur. Ophthalmological symptoms include repeated blurring of vision, progressive deterioration of vision, and cataract formation.

The course of the circulation, with special reference to the production of the symptoms, is described. Reference is made to a rise in the arterial tension in the lower limbs in certain cases.

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OBSERVATIONS ON THE BEHAVIOUR OF ERYTHROBLASTS CULTURED IN NORMAL AND "PERNICIOUS ANAEMIA" SERA

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Attempts to account for the action of Vitamin B₁₂ and folic acid in megaloblastic anaemias have led to reconsideration of the classical theory of the aetiology of pernicious anaemia. One of the new hypotheses is that advanced by Lajtha (1950), based largely on the results of marrow culture experiments. He confirmed the observation of Osgood and Brownlee (1937) that if megaloblastic bone marrow from a patient with pernicious anaemia in relapse is cultured the megaloblasts disappear more rapidly when the medium contains normal serum than when it contains serum from an untreated case of pernicious anaemia. He set up cultures with media containing different proportions of pernicious anaemia serum, and considered that the figures for differential counts showed that more megaloblasts persisted in the medium containing the higher proportion of pernicious anaemia serum. Lajtha therefore suggested that there exists in the blood of patients with pernicious anaemia a factor inhibiting the maturation of megaloblasts into either erythrocytes or normoblasts. Thompson (1950) has published evidence in favour of this suggestion.

The method employed by these workers is a modification of that described by Osgood and Brownlee (1937), using a fluid culture medium: this has the great advantage over methods employing a solid medium in that

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