Oclusion of the great vessels as they arise from the aortic arch has incited much interest among the medical profession, as evidenced by the numerous articles on the subject during the past decade. The nomenclature, clinical pictures, etiology, pathology, associated conditions, and treatment continue to be controversial. Credit for the initial clinical and pathologic descriptions of various forms of the condition remains questionable. Time and more knowledge of the pathophysiology of vascular disease have resolved some of the controversy. Yet, much remains enigmatic. Considerable confusion exists because different anatomic and pathologic conditions are capable of producing similar signs and symptoms. Classification has been so careless and complex that often a condition is labeled erroneously or a group of entirely unrelated conditions is discussed as the same entity. In this paper, an attempt will be made to clarify many of the discrepancies and to propose a practical classification and proper treatment.

**History**

In 1827, Adams' reported a case of a patient who had no peripheral pulses. This is considered by some, as the first description of aortic arch syndrome. Judge and associates refuted this claim, however, on the logical grounds that the clinical picture could represent cardiac shock, and necropsy revealed aortic stenosis and coronary artery disease. No mention was made of occlusion of any of the great vessels which arise from the aortic arch. In 1839, Davy and Dupuytren independently reported cases of patients with clinical manifestations of occlusion of the brachiocephalic vessels. Davy described the clinical course and postmortem observations in two patients. One had complete occlusion of all branches of the arch due to aneurysm of the arch. The other had occlusion of the left carotid and subclavian arteries by an unknown process, but the state of the innominate artery was not mentioned. Dupuytren's patient had gangrene of the nose, cheeks, and one little finger; however, pulses were reported as intermittent. At necropsy, the entire thoracic aorta, innominate, right subclavian, and both common carotid arteries were diffusely involved with bony patches, but neither occlusion nor severe stenosis of these vessels is mentioned. It is possible that peripheral ischemia was secondary to embolism or occlusive distal arterial disease. There is no conclusive evidence that Dupuytren's patient had occlusion of the great vessels of the aortic arch, whereas both of Davy's patients had occlusive vascular disease of the great vessels of the aortic arch, and therefore, his cases represent the first reports of aortic arch syndrome.

In 1908, Takayasu' described the case of a young woman with ocular signs which subsequently were known to occur with diffuse involvement of the brachiocephalic vessels. He made no reference, however, to obstruction of the great vessels. Onishi' recalled the absence of radial pulses in a patient who showed similar changes in the central retinal vessels.

In 1856, Savory' reported the case of a 22-year-old woman who had died with clinical evidence of brachiocephalic ischemia. The necropsy report read: "All the main arteries of both upper extremities and of the left side of the neck were reduced to..."
solid cords and presented the exact condition of vessels through which the flow of blood had been for some time mechanically arrested."

He described gross thickening of all three layers of the vessel wall. This we believe to be the first reported case of occlusion of the vessels of the aortic arch as the result of this peculiar form of arteritis, "Takayasu's arteritis."

Frøvig first suggested the term "aortic arch syndrome," which has been accepted as a most suitable term to encompass the whole spectrum of clinical and pathologic conditions. By repeated communications, Martorell and Fabre kept the medical profession aware of the presence of, and symptoms referable to, occlusion of the branches of the aortic arch. Informative reviews by Thurlbeck and Currens, Judge and co-workers, Ross and McKusick, and Ask-Upmark and Fajers have helped to clarify many variances and at the same time stimulate new conceptions of various aspects of the condition.

**Nomenclature**

Many terms have been used to designate occlusion of the vessel which arises from the aortic arch. These terms have described specific anatomic or pathologic conditions, etiologic factors, and clinical manifestations, or they have been named for a person who has contributed to our knowledge of the condition. Most of the terms may be applied accurately to a specific patient or group of patients, but unfortunately, they are often used to describe unrelated conditions. A general term may be used to include conditions unrelated except for one single anatomic, pathologic, or etiologic factor.

Among the names used to signify occlusion of the great vessels of the aortic arch are Takayasu's arteritis, aortic arch syndrome, obliteration of the supra-aortic trunks, thromboarteritis obliterans subclaviocarotica, panarteritis brachiocephalica, Takayasu's disease, Takayasu's syndrome, Martorell's syndrome, trunco-arteritis productive granulomatosa, pulseless disease, aortic arch arteritis, chronic subclavian-carotid obliteration syndrome, pulseless syndrome, reversed coarctation, brachiocephalic arteritis, branchial arteritis, and young female arteritis. Some of these titles indicate a particular cause, whereas others describe clinical manifestations, the location of obstruction, or the pathologic changes.

Aortic arch syndrome is a suitable term to denote a particular clinical picture produced by a condition involving the aortic arch. Although the term adequately describes the general syndrome, it does not cover a certain group in which obstruction of the brachiocephalic vessels distal to the aortic arch may produce the same clinical picture as that of occlusion or stenosis at the origin of the great vessels. The term gives no information about the clinical picture or cause of obstruction. Through the years, aortic arch syndrome has come to be associated with certain etiologic factors, such as arteriosclerosis or syphilis, but has not included obstruction caused by arteritis. Other terms have appeared in medical reports which signify a particular condition. Since Takayasu's description of the ocular changes, many Japanese physicians extended his study by investigation of various aspects of this condition. They have retained Takayasu's name to connote an arteritis that causes obstruction of the branches of the aortic arch in young females. Today "Takayasu's arteritis" is specific for this condition, but it has no relation to occlusion of the brachiocephalic vessels from other causes.

It appears to us that a general term which signifies obstruction of the vessels of the aortic arch and their branches should be used to name this condition. Such a term as "brachiocephalic ischemia" would cover occlusion or stenosis in any of the brachiocephalic vessels, regardless of the cause or location. To this could be added subtitles indicating cause and degree and location of obstruction. For example, brachiocephalic ischemia due to arteriosclerotic occlusion of the innominate artery, stenosis...
of the left common carotid artery, and no significant disease in the left subclavian artery would then be classified as Brachiocephalic Ischemia.

A. Arteriosclerosis.
B. Occlusion, innominate artery stenosis, left common carotid artery.

Omission of mention of a vessel would imply patency of this vessel. This classification should be further extended to include obstruction of the vessels distal to their origin and their branches. Simple occlusion of a carotid, vertebral, or subclavian artery could still be classified as brachiocephalic ischemia, although occlusion of a single vessel could be designated as either brachial or cephalic ischemia. Single involvement of the innominate or either subclavian artery proximal to the origin of the vertebral should be designated brachiocephalic ischemia because such occlusion produces both brachial and cephalic ischemia.

The clinical manifestations of the aortic arch syndrome (brachiocephalic ischemia) need not be the result of occlusion of the great vessels of the aortic arch at their origin. Obstruction distal to their origin or occlusion in several of their branches can produce the classic clinical picture associated with obstruction of all the great vessels at their origin if there are enough sites of distal obstruction (Fig. 1). A clinical picture typical of what is considered an aortic arch syndrome may be produced when the aortic arch is entirely uninvolved. Therefore, brachiocephalic ischemia best describes the condition, regardless of the cause or site of vascular obstruction.

**ETIOLOGY**

Any pathologic process, whether intrinsic or extrinsic, capable of occluding a blood vessel can cause brachiocephalic ischemia. The principal etiologic factor in this country undoubtedly is arteriosclerosis and in the Middle and Far East, an obscure arteritis (Takayasu's arteritis). Most reported cases of occlusion of the branches of the aortic arch are due to these two causes. Reports of Takayasu's arteritis have been particularly prolific because of the ambiguous nature of the disease, interesting clinical picture, high frequency of associated conditions, and wide speculation regarding its true nature. Conditions which may cause

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Image: Figure 1: A. Archogram of patient with manifestations of brachiocephalic ischemia. Note normal patency of vessels at their origin from arch of aorta, and multiple areas of stenosis distally. B. Drawing of major vessels, as seen in archogram. Arrows point to areas of stenosis.
occlusion of the branches of the aortic arch are: arteriosclerosis, Takayasu's arteritis, syphilitic aortitis, aneurysms of the aortic arch, dissecting aneurysm, thromboangiitis, traumatic thrombosis, tuberculous arteritis, allergic arteritis, giant cell arteritis, embolism, congenital anomalies, thoracic outlet syndrome, and thrombocytosis.

Arteriosclerosis is a generalized arterial disease with a predilection for certain sites in the cardiovascular system. It is the most common occlusive disease of the aortic arch and its branches. Many other parts of the circulatory system may be affected alone or concomitantly. Although all branches of the aortic arch may contain atheromatosus plaques, rarely will all the branches be completely occluded. When arteriosclerosis produces obstruction of the aortic arch, distal sites often are likewise involved, particularly the bifurcation of the carotid arteries and at the origin of the vertebral arteries. Arteriosclerosis accompanies various types of arteritis in their later stages and frequently is the actual cause of arterial occlusion. Certain pathologic and physical conditions, such as syphilis, injuries, and stress, make the aortic arch more vulnerable to athromatosis.

Takayasu's arteritis generally occurs in women of childbearing age and at one time was thought to be peculiar to Asiatic people. Various forms of peripheral and central arteritis are more common than generally believed. The site of obstructive disease determines the clinical picture. Despite numerous clinical and pathologic studies, the cause of Takayasu's arteritis remains obscure, but its association with a number of conditions has led to considerable speculation. Arteritis is the most common pathologic alteration at one end of the broad spectrum of collagen disorders, and since the systemic manifestations of many collagen diseases are common in patients with Takayasu's arteritis, some have postulated a relationship between the two diseases. An autoimmune mechanism in Takayasu's arteritis is further supported by its reported association with rheumatoid arthritis, polymyositis, and ankylosing spondylitis. Also, the histologic vascular changes in Takayasu's arteritis are similar to those seen in autoimmune diseases. Positive LE cell preparations and the electrophoretic pattern in patients with Takayasu's disease have been the subject of much debate. Although many characteristics of Takayasu's arteritis suggest an autoimmune cause, this etiologic theory is difficult to support clinically and pathologically in every case. On the other hand, it is also difficult to exclude the importance of such a process in specific cases. Isolation of infectious organisms, such as Treponema or Streptococcus, has proved futile. Hemodynamic stress, endocrine imbalance, hypersensitivity phenomena, collagen disorders, and allergic reactions have all been incriminated, but none has been definitely proved.

Undoubtedly, other causes besides arteriosclerosis and Takayasu's arteritis are responsible for brachiocephalic ischemia. Ross and McKusick showed syphilitic aortitis, with or without aneurysm, to be a major cause. Dissecting and traumatic aneurysms also obstruct the vessels, but in such instances the clinical picture is acute and the obstructing mechanism easily diagnosed. Other causes, listed earlier in this paper, have been reported or are theoretically capable of producing the classic picture.

A variant of Takayasu's arteritis has been reported in children. Paton and co-workers recorded seven cases of aortic arteritis in children from Thailand, aged four to 12 years. Warshaw and Spach reported a case of arteritis of the aortic arch in a seven-year-old girl.

Occurrence of Takayasu's arteritis and Hodgkin's disease in the same patient has been reported twice. In one patient, brachiocephalic ischemia existed before radiotherapy for Hodgkin's disease and in the other patient, ischemic manifestations developed after radiation therapy. The first case probably represents a coincidence, whereas the second is probably due to radiation injury.
AORTIC ARCH SYNDROME

CLINICAL PICTURE

The clinical manifestations of brachiocephalic ischemia depend on the vessels involved, the degree of obstruction, and the systemic manifestations associated with a particular disease. The stage and rapidity of vascular occlusion, together with the extent of development of collateral circulation, also influence the degree of vascular insufficiency. In general, the predominant or presenting complaint is referable to cerebrovascular insufficiency. Rarely is the patient's major concern about disturbances in the arms. Occlusive disease of the cerebrovascular system is usually preceded by transient focal cerebral ischemic attacks ("little strokes"). Symptoms range from a minor neurologic deficit to those of full-blown stroke. These "little strokes" last from a few seconds to hours, but a series may occur during the succeeding weeks or even months. Although usually normal function returns, in many the disease progresses to cerebral infarction. The various manifestations of cerebrovascular insufficiency may be difficult to analyze in attempts to localize the area of arterial stenosis or occlusion. If many segments of a vessel are involved, localization may be particularly difficult. Reasonable diagnostic accuracy, however, can be obtained, since a particular arterial distribution is manifested clinically in a specific manner. Carotid artery insufficiency is characterized by ipsilateral blindness, contralateral hemiplegia or hemi-anesthesia, confusion, headache, and if the occlusion is on the dominant side, dysphagia or aphasia. Insufficiency of the vertebral artery shows considerably more variation in the number and nature of symptoms. Confusion, blindness, dysphagia, vertigo, dysarthria, ataxia, diplopia, visual field defects, and numbness or weakness on one side of the face or body are among the common symptoms of vertebral-basilar ischemia.

Martorell and Fabre presented the classic description of a patient with chronic occlusion of all the great vessels at their origin from the aortic arch. The features of this syndrome are facial atrophy; orthostatic syncope and epileptiform attacks; cranio-cervical pain, especially during mastication; transient visual disturbances; cataracts; weakness and paresthesias of the arms, and progressive loss of weight. Although the description is classic, the features are more characteristic of occlusive disease of the aortic arch caused by arteritis than by arteriosclerosis. In the former, signs and symptoms are due to impoverished blood supply to the entire head and upper limbs, whereas in the latter they are related to diminished blood supply to the brain and occasionally the upper limbs. The difference occurs because frequently all great vessels are occluded in the patient with arteritis, life being maintained only by collateral circulation which was able to develop because the occlusion was chronically progressive. Arteriosclerosis, on the other hand, rarely causes complete occlusion of all vessels of the aortic arch, although complete occlusion of a single vessel at the aortic arch and stenoses of the other vessels do occur. Arteriosclerotic occlusion is usually restricted to the left subclavian or innominate artery alone.

Vertigo, syncope, transient visual disturbances, and headaches were the most frequent neurologic manifestations in three large reported series of aortic arch syndrome. Most of the patients had occlusion of all of the great vessels by Takayasu's arteritis, although other causes were incriminated in a minor number.

Ophthalmologists have studied extensively the various ocular manifestations in brachiocephalic ischemia, since many patients first seek medical advice because of visual disturbances. Particular attention has been paid to the changes in Takayasu's arteritis. Pinkham listed in order of frequency the following abnormal ocular changes: transient visual disturbances, 70 per cent; cataracts, 45 per cent; retinal arteriovenous anastomoses, 39 per cent; atrophy of the iris, 38 per cent; retinal hemorrhage, 38 per cent; micro-aneurysms, 33 per cent; mydriasis, 32 per cent; hyperemia
of conjunctive and sclerae, 26 per cent; dilated retinal vessels, 23 per cent; retinal detachment, 20 per cent; and exophthalmos, 18 per cent. Measurement of the central retinal arterial pressure by ophthalmodynamometry has been useful in determining the status of carotid circulation, although this method is by no means flawless, since both false positive and negative results are common. Peripheral obstruction and collateral circulation may lead to an erroneous interpretation.

Hypersensitive carotid sinuses associated with Takayasu's arteritis have been frequently described. The active role of a carotid sinus in cerebrovascular insufficiency is supported by report of a patient with carotid sinus symptoms after ligation of the common and internal carotid arteries, and relief of such symptoms after surgical removal of the carotid bifurcation. In patients with cerebrovascular insufficiency, there is often a delicate balance in the cerebral blood flow, and any further reduction, no matter how infinitesimal, may elicit symptoms. Those experienced in the examination of patients with cerebrovascular insufficiency are aware of the frequency with which symptoms can be produced by minor pressure on a carotid bifurcation of the predominant vessel. It seems logical to assume that the symptoms referable to hyperactive carotid sinuses are in reality due to further reduction in cerebral blood flow, caused by tactile pressure.

Absence of pulsations in the neck and arm is an essential part of brachiocephalic ischemia, as indicated by the term "pulseless disease." Also included in brachiocephalic ischemia are patients with occlusion of only one or two of the three great vessels of the aortic arch. The magnitude of the pulse is limited to the degree and extent of vascular involvement, and often the pulse is diminished simply as the result of stenosis. Pulses in the arm become impalpable before the blood pressure becomes completely unattainable by Korotoff sounds. Although the pulse pressure may be greatly reduced, the mean blood pressure is usually only slightly reduced, being maintained by collateral circulation.

Murmurs in the supraclavicular region usually denote stenosis of a vessel. The presence of a murmur, however, does not necessarily mean a decrease in flow or pressure. Distal arterial pressure and flow are not affected until the degree of stenosis becomes critical (75 to 80 per cent of the luminal area), after which minute increases in stenosis cause precipitous reduction in distal flow and pressure. Tortuosity without narrowing of the vessel may also produce a murmur.

Facial atrophy, which has been described as a feature of brachiocephalic ischemia, is due to a greatly decreased blood supply to the facial muscles. Since there is a vast network of collateral vessels in the face, neck, and shoulders, it is difficult to deplete the muscles of the face of their blood supply. We have not seen patients with facial atrophy in which the ischemia was secondary to arteriosclerosis, but vessels are often occluded by arteritis. Trophic changes in the hand are rare. In Takayasu's arteritis, trophic changes of the head and face, such as necrosis of the nose, palate, and lip; alopecia; and perforated nasal septom, have been reported. Frequently associated with Takayasu's arteritis are systemic manifestations (fever, fatigue, headache, and arthralgia), and consistent abnormal results of laboratory studies (anemia, leukocytosis, and elevated erythrocytic sedimentation rate). The frequency of these systemic manifestations has led to the postulation that many cases of brachiocephalic ischemia are the result of a generalized process, be it a rheumatoid, infectious, or auto-immune reaction.

Hypertension is common in patients with brachiocephalic ischemia. When pressure is unobtainable in the arms, hypertension in the leg is common. Ask-Upmark found hypertension in 50 per cent of patients with Takayasu's arteritis. The elevated blood pressure in these patients does not seem to be the result of mechanical obstruction because diastolic pressures also are elevated.
Defective cerebral circulation has been suspected as the etiologic factor in hypertension, since Fishback and associates were able to produce hypertension experimentally in dogs by progressive ligation of the arteries supplying the head. The high incidence of associated stenosing lesions in the renal arteries has led to the conclusion that hypertension is probably of renal origin in most cases of brachiocephalic ischemia. All the etiologic factors which produce changes in the brachiocephalic vessels are likewise capable of producing stenosis of the renal artery.

Recently, a particular form of brachiocephalic ischemia has received much attention. When proximal occlusion of the subclavian or innominate artery produces centrifugal blood flow in the vertebral artery of the involved side, the condition is known as "subclavian steal." A distant, patent subclavian artery drains blood via the vertebral artery from the circulation of the brain (Fig. 2). Evidence of cerebral ischemia may not be manifest unless other portions of the cerebral circulation are diminished. The clinical picture is that of vertebral-basilar arterial insufficiency, which is exaggerated by exercise of the arm.

Pathology

The pathologic changes caused by brachiocephalic ischemia depend on the cause of obstruction. Of the two common causes, perivascular involvement appears less frequent in arteriosclerosis than in Takayasu's arteritis. Cohen and co-workers suggested that a differential point in histologic diagnosis is that a thickened mantle of perivascular connective tissue is present in arteritis, but absent in arteriosclerosis. This is not always the case, however, for perivascular reaction is occasionally noted in arteriosclerosis. The atheromatous intimal plaque of arteriosclerosis is characterized by focal deposition of lipids and fibrosis in the subendothelial connective tissue of the in-
The ulcerated lumen. The plaque may progress to fibrosis or calcification and even ulcerate into the lumen. It is this plaque, or more often the ulcerated plaque, which predisposes to mural thrombosis, leading to severe deformity, narrowing, and eventual occlusion of the arteries. In arteritis the morphologic changes depend on the stage of the process. The pathologic changes usually involve all the elastic arteries of the aortic arch and appear to begin in the adventitial coat. The cellular reaction is intense and subsequently involves the media and the intima. Reaction is characterized by infiltration with leukocytes, chiefly lymphocytes and plasma cells, with eventual destruction of the elastic lamina and muscle fibers and subsequent replacement by connective tissue. The early stage of arteritis can easily be differentiated from arteriosclerosis, but as the disease progresses with long duration and superimposed degenerative changes, differentiation may be difficult.

In both conditions, the distal portion of the involved vessels is spared to a varying degree. This is particularly true in Takayasu's arteritis, whereas in arteriosclerosis, distal sites sometimes are involved, especially at points of maximal stress, such as branchings and areas of fixation of the vessels. In arteriosclerosis, the abdominal aorta and its branches are often also involved; consequently renovascular hypertension and occlusive vascular disease of the legs are common. Although at one time thought to be rare, involvement of other vascular sites in Takayasu's arteritis appears rather common. Atypical coarctation of the aorta and renal stenosis have been frequently reported.

**Treatment**

A stenosing or obstructing lesion of the branches of the aortic arch does not necessarily imply brachiocephalic ischemia. The congenital and acquired status of the collateral circulation to the brain determines the degree and type of neurologic manifestations. Adequate collateral circulation is most apt to develop after progressive but slow occlusion of a vessel. Cerebral

![Figure 3: Direct archogram via percutaneous transfemoral retrograde catheterization. Archogram demonstrates occlusive involvement of all vessels at their origin from the aortic arch.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21450/)

![Figure 4: Operative photograph of aorto-bilateral carotid bypass, with bifurcated Dacron prosthesis. Arrows point to anastomoses. Note the early bifurcation of the prosthesis and lateral placement of the limbs.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21450/)
communications in the circle of Willis and through the external carotid artery are of prime importance, but blood pressure and cardiac output also play a significant role. There is uniform agreement that symptomatic, extracranial, vascular occlusion demands urgent surgical treatment. Controversy, however, still exists regarding the value of prophylactic surgical treatment of asymptomatic, angiographically demonstrable, extracranial arterial lesions. In an interesting study by Faris and associates,\textsuperscript{44} angiographic visualization of all four extracranial cerebral arteries in 43 asymptomatic volunteer inmates at Kansas State Penitentiary revealed 53.6 per cent had either intrinsic or extrinsic arterial abnormalities, and 23.2 per cent had involvement of more than one artery. The abnormalities were due to intravascular plaques, presumably arteriosclerotic, in 53 per cent of the patients. In 5 per cent, one vessel was completely occluded. Undoubtedly, many persons with varying degrees of occlusive cerebrovascular disease will live a complete life span without symptoms. It would be absurd to subject patients with minor degrees of arterial stenosis to operation. Yet, it seems wise to correct asymptomatic severe stenosis (85 per cent or greater) to prevent a possible catastrophe.

Most agree that asymptomatic brachial occlusive lesions do not require surgical intervention. Treatment of symptomatic brachial lesions depends on the severity of symptoms, plus the functional desires of the patient.

If symptoms of brachiocephalic ischemia are due to segmental occlusion, the logical approach is operative, that is, bypass grafts or thromboendarterectomy, with or without enlargement by a patch graft. In Takayasu's arteritis, encouraging results in controlling the disease with steroids have been reported.\textsuperscript{45} Here, the goal is to stabilize the disease, relief of symptoms being unlikely. The only other form of nonoperative therapy is anticoagulation,\textsuperscript{46} which has occasionally yielded favorable results in patients unsuitable for surgical correction.

The type and extent of surgical correction must be individualized. The general condition, severity of symptoms, extent and type of obstructing vascular lesion, and socio-economic status of the patient govern the choice of operative procedure. Many surgical technics have been used, and a particular physiologic or anatomic situa-

\textbf{Figure 5:} Drawing depicts aorto-subclavian bypass of occluded innominate artery. Vertebral and carotid arteries fill by retrograde flow.
tion may be ideal for a specific operation. Usually, the vessels distal to obstruction are patent, and reconstruction is possible with restoration of normal hemodynamics. Occasionally, there may be concomitant localized distal arterial obstruction, which also is amenable to surgical correction.

Aortography is the ideal method of delineating the extent and nature of vascular involvement. Direct archogram via retrograde cannulation provides the best visualization of the aortic arch and its branches. Injection of a 60 ml bolus of sodium diatrizoate (90 per cent Hypaque) with 190 sq lb of pressure has proved practical and safe. The percutaneous transfemoral retrograde route by the Seldinger technic is preferable” (Fig. 3). In some cases, arteriosclerotic obstruction of the iliac and femoral vessels makes retrograde cannulation dangerous or impossible. If the right subclavian and innominate arteries are patent, equally satisfactory direct archograms can be obtained via a retrograde right brachial route. If the patient’s body is positioned and the head rotated, all the vessels as they arise from the arch, as well as their distal branches, can be adequately visualized by serial exposures. If retrograde catheterization is impractical, venous arch angiography must be used, but delineation of the vascular structures is somewhat limited.

Various operative approaches have been suggested, but a median sternotomy incision gives the best exposure of the diseased vessel, and also is a suitable approach for bypass procedures. The incision can be carried to the diseased side of the neck, and if both sides are affected, a small counter-incision can be made for exposure.

The choice of operative technic usually lies between endarterectomy and bypass graft. In occlusion of the great vessels at their origin, adequate endarterectomy is often difficult and fraught with some danger. Proximal endarterectomy requires partial occlusion of the aortic arch with limited exposure and hazards of producing a plane of dissection within the aortic wall. Bypass grafts allow one to choose the least involved portion of the aorta and select a distal site which is accessible and suitable for anastomosis. Bypass also has the advantage of simultaneous treatment of two or more vessels and bypass of multiple areas in a single vessel. If two or more branches

![Figure 6: Drawing of aorto-bilateral carotid bypass of occlusion of the innominate and left common carotid arteries at their origin, and stenosis of right subclavian artery at its origin. Right vertebral and right subclavian arteries fill by retrograde flow.](image-url)
of the arch are involved, we prefer use of
a graft which bifurcates within the thorax
(Fig. 4). Concern has been expressed over
such a graft producing obstruction of struc-
tures at the thoracic inlet. If a short cen-
tral portion is used, so that the graft bifur-
cates low in the mediastinum and its limbs
are brought into the neck laterally and be-
exthe innominate veins, there will be
no impingement on the thoracic inlet struc-
tures. Bypass graft should also be small, the
limbs being no more than 8 mm in diame-

**FIGURE 7**: Drawing shows occlusion of all branches at their origin from the aortic arch. A bifurcation
prosthesis is used to bypass the obstructed vessels. One limb is anastomosed to the right carotid artery
and the other limb is anastomosed first to the left carotid artery and then to the left subclavian artery.

**FIGURE 8**: Drawing shows various surgical technics used to correct subclavian steal. A. Endarterectomy
with patch graft. B. Aorto-subclavian bypass with autogenous vein. C. Carotid subclavian bypass with
Dacron prosthesis.
ter. If temporary occlusion of a stenosed carotid artery is necessary for distal anastomosis, an internal shunt should be used to insure adequate cerebral blood flow. When the origin of the innominate artery is occluded and there is adequate communication between the subclavian and carotid arteries, temporary occlusion of the carotid artery can be prevented by performing the distal anastomosis in the subclavian artery. Cerebral circulation is reestablished by retrograde flow to both the vertebral and carotid arteries (Fig. 5). Should communication between the subclavian and carotid arteries be inadequate, however, it is wiser to restore carotid flow (Fig. 6). When all three vessels are occluded at their origin, a bifurcation graft should be used to deliver blood from the central ascending aorta. One limb should be anastomosed to the right subclavian or carotid. The other limb should be anastomosed first side-to-side to the left carotid and then continued to the left subclavian as an end-to-side anastomosis (Fig. 7). This will restore flow to all brachiocephalic vessels, assuming there is patency between the branches of the innominate artery. Occlusion of a single vessel at its origin from the aortic arch occurs most frequently in the left subclavian. Here blood reaches the subclavian artery via the ipsilateral vertebral artery, siphoning blood from the cerebral circulation. This phenomenon, referred to as subclavian steal, can occur without production of neurologic symptoms. Often, another cerebrovascular lesion must be present before symptoms are provoked. We have encountered this phenomenon in 20 patients, but in only 12 did symptoms warrant operative correction. Various methods to correct this condition are shown in Fig. 8. If endarterectomy is feasible, a patch is used to widen the arteriotomy site. The occluded proximal subclavian artery may also be bypassed with a prosthetic or preferably a vein graft. Both these methods require a posterolateral thoracotomy incision. A less traumatic approach is to “jump” a graft from the common carotid artery to the subclavian artery, distal to the obstruction. This maneuver can be performed through a transverse cervical incision, but care should be taken to insure carotid flow during the proximal anastomosis. Although this procedure would appear to steal blood from the carotid circulation, symptoms of carotid insufficiency have not developed in four of our patients who had the operation. In essence, the operation transforms the common carotid artery to a brachiocephalic or innominate artery.

If more than one artery or branches of an artery are involved, selection of the artery or arteries of greatest importance to be reconstructed is determined by the degree of patency of the contralateral vessels, their intracerebral and extracerebral connections, and neurologic signs.

A word of caution is needed when associated lesions, such as atypical coarctation and renal arterial stenosis, cause hypertension. It might prove catastrophic to correct these lesions in the presence of brachiocephalic ischemia, for depression of blood pressure in the presence of impaired cerebral flow may produce cerebral infarction. Consequently, cerebrovascular obstruction should be relieved before correction of concomitant hypertensive vascular lesions.

**Summary**

Occlusion of the great vessels as they arise from the aortic arch has been designated in a variety of ways to describe the clinical manifestations, anatomic or pathologic alterations, etiologic factors, or the name of a person associated with the condition. Brachiocephalic ischemia best describes the result of obstruction of the branches of the aortic arch.

The exact clinical manifestations depend on the cause and anatomic distribution of the occlusive process, the coexistent systemic manifestations of the disease, and the status of the collateral circulation. It is not surprising, therefore, that the symptoms and course of brachiocephalic ischemia are extremely variable. Transient episodes of cerebral ischemia are produced, and it is mandatory to recognize these warning signs
so that surgical treatment can be instituted before irreversible damage occurs.

Surgical treatment is accomplished by bypass graft or thromboendarterectomy, with or without patch graft. Selection of the type and extent of surgical reconstruction must be individualized, and determined by the severity of symptoms, anatomic and pathologic nature of the disease, and physiologic status of the patient.

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AORTIC ARCH SYNDROME


For reprints, please write: Dr. Ochsner, 1514 Jefferson Highway, New Orleans 70121.

GIANT LEFT ATRIUM AS CAUSE OF DYSPHAGIA

Two cases have been presented showing the typical roentgenologic pattern of extrinsic pressure on the esophagus from the giant left atrium. Both patients complained of pain, progressive difficulty in swallowing and regurgitation of food; and both had temporary relief in symptoms with diuresis before surgery.


CARDIAC MOTION

Diagnostic ultrasound was used to record the movement of the anterior and posterior heart walls in 23 patients with proved pericardial effusion. Fifteen of these patients demonstrated heart wall motion that could be distinguished from the normal pattern. Two patients with acute cardiac tamponade exhibited markedly reduced cardiac motion. The ultrasound cardiograms in the remaining six patients indicated posterior displacement of the entire heart during systole. The extent of this cardiac displacement varied. Five of the six patients with exces-

sive cardiac motion had malignant pericardial effusion and four of the six had clinical evidence of cardiac tamponade. The results of this study substantiate the theory that excessive cardiac motion may occur in some patients with pericardial effusion. Objective evidence is also provided in support of the positional etiology of electrical alternation associated with pericardial effusion.