ABNORMAL arteriovenous communication, such as a septal defect or an arteriovenous fistula, is characterized by certain physiologic and pathologic phenomena that make it one of the most fascinating and unique lesions in medicine. Only an intimate knowledge of these phenomena, and their cause, permits a complete understanding of the many diverse effects of this extraordinary lesion upon the circulatory system, most particularly upon the heart.

In utero, communications between the arterial and venous circulations occur normally as an interatrial septal defect, and as the ductus arteriosus which circumvents the inactive lungs by transferring blood from the pulmonary artery to the descending aorta. Such communications (normal before birth) may persist after birth as abnormal communications of variable size and effect. Early in embryonic life, before circulation of blood has begun, there appears in the course of development a defect in the interventricular septum, which though serving no normal function at the time, persists into later intrauterine life and remains after birth as an abnormal arteriovenous communication. Also, in the embryonic peripheral bed, vessels which have served as arteries may eventually become veins, and veins may ultimately serve as arteries. "The fact that arteries and veins develop out of a common capillary plexus forms the basis for the persistence of direct connections between them." (Sabin.)

Acquired communications between the two systems occur as arterial and venous aneurysmal dilatations with a connecting passage between them, the so-called "varicose aneurysm," or as direct anastomoses produced by gunshot or stab wounds, by inclusion at operation of artery and vein in the same ligature, or by erosion of the contiguous walls of the two vessels by blood-borne infecting agents.

These congenital and acquired fistulas, if sufficiently large, have several remarkable characteristics that are common to both. Under no other circumstances is it possible to produce such a startling rise in general blood pressure, followed immediately by an equally startling slowing of the heart beat, as may be produced by closing a large peripheral fistula or by blocking the artery proximal to a large, widespread congenital hemangioma. To explain these unusual phenomena, an increase in the total volume of circulating blood was postulated, a concept not previously proved in circulatory disorders. This explanation was immediately, and subsequently, challenged by other students of this lesion, challenges that stimulated the performance of numerous experiments to determine the effects of arteriovenous fistulas of various sizes, in various locations.

Application of these experimental studies to clinical cases of arteriovenous communica-
tions, including the congenital aberrations, permitted an elaboration and subsequent verification of acceptable explanations for some of the unusual and puzzling features of this entity. To the underlying theoretic pathologic physiology as presented in 1923 were added important ancillary concepts in subsequent presentations.12-15

Of particular significance is the concept that an abnormal communication in the circulatory system, wherever located, establishes two circuits in which the blood, though flowing in part through the same conduits, may be visualized as flowing through two separate systems, one characterized by high arterial pressure and high peripheral resistance, and a second system bypassing part of the first and thus introducing a shorter circuit, characterized by low pressure and low peripheral resistance, each supplied with flowing blood from the same central source, the heart. Obeying the law of hydraulics that flowing blood like flowing water seeks the path of least resistance, such a communication, whether peripheral or intracardiac, whether acquired or congenital, provides an ever-present, irresistible avenue for the escape of blood from the system of high pressure and high resistance into the capacious shorter system of low pressure and low resistance. This shorter circuit serves no useful purpose, and is in fact a parasitic circuit engraven upon the normal circulation and capable of producing serious deleterious effects.16

A clearer conception of the flow through the two circuits may be obtained from the highly schematic diagram in figure 1. In the presence of an abnormal opening, FF¹, between circuit A of high capillary resistance and circuit B of little or no resistance, blood will flow directly from circuit A into circuit B without traversing a capillary bed, the volume depending on the size of the opening and the resistance to flow imposed by the fistula at FF¹ and by the limited distensibility of the artery leading to it. In the presence of a small fistula, or of a moderate fistula with firm, fibrous borders, the volume flow through FF¹ is minimal, and the dilatation of the components of the fistula circuit AFB will be minimal. A small fistula may, and frequently does, close spontaneously through contraction of the fibrous tissue forming the rim of the opening. A fistula or intracardiac shunt, small or moderate in size with rigid borders, may remain stationary in size with no demonstrable deleterious hemodynamic effects upon the heart or upon the circulation except for the thrill and bruit at the site of communication. If, however, the rim of the fistula is elastic, distensible, and, therefore, capable of yielding to the high arterial pressure of circuit A by progressive expansion, the volume or bulk of blood transmitted through the fistula and sequestered in the fistula circuit may increase progressively, causing progressive dilatation of all the components of the fistula circuit.

The evidence for this progressive dilatation of the fistula in a few cases, upon which the occasional remarkable progressive dilatation of the remaining components of the fistula circuit depends, is unassailable. For example, in four personally treated femoral fistulas of 25,2 26,12 24,10 and 1517 years' duration, cardiac dilatation progressed slowly through these many years to complete cardiac disability, accompanied by marked dilatation also of the artery proximal to the fistula. The
fistulas at operation were found to have diameters of 2.1, 1.8, 1.4, and 2.5 cm., located in arteries whose measured diameters proximal to the fistula were 1.8, 2.1, 2.1, and 2.8 cm., respectively, diminishing immediately distal to the fistula to only 0.8, 0.7, 0.65, and 0.7 cm., respectively. The normal femoral arteries in the opposite extremity had diameters by palpation of only 0.6, 1.0, 1.0 cm., respectively (the diameter of the normal opposite artery in the fourth case was not recorded). Obviously, a fistula 2.0 cm. in diameter would be difficult if not impossible to produce at the initial injury in a vessel only 0.6 cm. in diameter.

An important attribute of this progressive expansion of the occasional fistula is its fortuitous and erratic development, since this depends so intimately upon the vagaries of the healing process in the course of which fibrous tissue of greater or less extent is deposited around the fistula or around one or more of the four orifices of the two vessels between which the fistula lies. The constriction of each one of these orifices has its own specific effect upon the flow through the fistula, as proved experimentally.\(^\text{18-20}\) Moreover, all gradations in the extent and distensibility of these deposits of fibrous tissue may occur, affecting critically the occurrence, the extent, and the rapidity of the dilatation of the fistula, and, therefore, also of the dilatation of the heart and of the artery leading to the fistula.

Equally important is the concept that the progressive expansion of a fistula or of a congenital defect, can be insidiously slow, with only infinitesimal increments—perhaps as little at a time as 2 or 3 mm.\(^\text{8}\) of added blood being forced through the abnormal opening into the circuit of low resistance, probably most often during moments of circulatory stress, and repeated endlessly in the course of the years during which dilatation of the fistula and of the heart develops. Once an additional quantity of blood has been forced through the fistula, the volume of blood flowing through the fistula circuit will be augmented permanently by this amount, and the components of the fistula circuit will be dilated permanently to an equivalent degree, no matter how infinitesimal the increment may be.

It is this inevitable shunting and sequestration of a greater and greater volume of blood into the shorter circuit of low resistance and low pressure through a progressively but slowly expanding fistula that provides the long-sought explanation for the remarkably protracted cardiac dilatation and ultimate cardiac failure that developed so insidiously in the presence of fistulas of 25, 26, 24, and 15 years' duration, as described above, and in the presence of fistulas of 48 and 57 years' duration as reported by Dinak\(^\text{21}\) and Dorney,\(^\text{22}\) respectively. Accompanying this dilatation of the heart was a dilatation of all components of the fistula circuit, including more particularly a marked dilatation of the artery proximal to the fistula.

It should be noted that although this cardiac dilatation may not occur at all, it may also occur very rapidly, as observed by Mason\(^\text{23}\) in a patient with a traumatic left subclavian-left innominate vein anastomosis which produced cardiac failure in 2\% months, completely controlled by elimination of the fistula. Much depends upon the ease with which blood can be transferred by the high pressure of the normal circuit into the circuit of low resistance through an easily expansible fistula. In Mason's case the negative pressure, which so frequently occurs normally in the innominate vein, and the high pressure of the subclavian artery, as well as an easily dilatable fistula, undoubtedly contributed to the rapid development of cardiac dilatation and failure. Experimentally, cardiac failure occurred within 3 weeks in the presence of an aorto-vena cava fistula with prompt regression following elimination of the fistula.\(^\text{9}\)

Associated with these striking features of a peripheral arteriovenous fistula are an increased venous pressure not only distal to the fistula, but also in the vein and vena cava proximal to the fistula commensurate with the volume flow through the fistula,\(^\text{15}\) and an increase in cardiac output commensurate with the size of the fistula and with the velocity
of flow through it.\textsuperscript{2, 18, 24} The site of greatest velocity of flow in the circulatory bed lies in the ascending aorta, both velocity and size of the aorta diminishing rapidly toward the periphery as the number of primary and secondary branches of the aorta increases, and their sizes diminish. Hence the velocity of flow through an aorta-vena cava fistula is greater than through an iliac fistula of equal size, and greater through an iliac fistula than through a femoral fistula of equal size, thus accounting in part for the differing effects of these differently located fistulas. A remarkable cardiac output of 17.2 liters was demonstrated recently by cardiac catheterization in the presence of a large iliac fistula of 17 years’ duration accompanied by great dilatation of the heart and proximal artery: 9.6 liters through the fistula circuit and 7.6 liters through the normal systemic circuit.\textsuperscript{25} Binak\textsuperscript{21} demonstrated an even more remarkable cardiac output of 23.9 liters per minute in the presence of a femoral fistula of 12 years’ duration which had produced a 49-per cent increase in the size of the heart.

This remarkable latent capacity of a heart to accept over the years a greater and greater volume load of flowing blood can be explained, first, by the low peripheral resistance of the fistula circuit against which the heart must work in propelling forward the increased volume of blood, and, second, by a concomitant hypertrophy of the cardiac musculature in response to the increased volume flow, which produces a commensurate increase in cardiac work. This increased cardiac work in addition to the marked cardiac dilatation is undoubtedly an important factor in the cardiac failure that ultimately occurs so conspicuously in some cases. The extent of such hypertrophy was well illustrated in one of our experiments\textsuperscript{15} in which two puppy littermates were subjected to aorta-vena cava fistulas, a third puppy of the same litter being kept as control. At necropsy a year later, the control puppy’s heart weighed 74 Gm.; the heart of pup 1 with a fistula 12 mm. in circumference, as measured at necropsy, weighed 123 Gm.; and the heart of pup 2 with a fistula 18 mm. in circumference weighed 176 Gm. (fig. 2). The aorta and vena cava proximal to the fistula were dilated commensurate with the size of the fistula, greater in pup 2. At the time of necropsy, the three animals were in good condition without signs of cardiac failure, free from edema and ascites.

As a direct corollary to the sudden diversion of blood into a fistula circuit is an immediate reduction in the volume of blood flowing in the normal circuit with a corresponding reduction in the arterial pressure of this circuit. An immediate compensation for the diverted blood occurs through accesses to the total volume of circulating blood from the normal reservoirs in the body: the liver, the spleen, and the splanchnic area; followed by a progressive permanent increase in total blood volume paralleling more or less closely the increasing volume flow through the fistula circuit.\textsuperscript{18} The total blood volume is probably increased by the same bodily mechanisms responsible for restoration of blood volume following an external hemor-

\textbf{Figure 2}

Cross-section of hearts of 3 litter mates: (a) heart of control animal weighed 74 Gm.; the animal weighed 11.6 Kg.; (b) heart of animal with aorta-vena cava fistula 12 mm. in circumference weighed 123 Gm.; the animal weighed 10 Kg.; (c) heart of animal with aorta-vena cava fistula 18 mm. in circumference weighed 176 Gm.; animal weighed 11.1 Kg. The cardiac dilatation and hypertrophy were commensurate with the size of the fistulas, which were of 1 year’s duration.

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rhage massive enough to lower blood pressure. On operative closure of the shunt, this blood volume increase is directed into the normal circulatory bed, distending it and causing a prompt elevation in general arterial pressure, which is promptly though only partially rectified by a slowing of the heart beat initiated by stimulation of the vagus in the abnormally distended aortic arch and by abnormal distention of the cerebral vessels.\(^{26}\) Atropine abolishes this retardation in pulse rate, indicating its vagal origin. Atropine has no effect, however, upon the elevation of blood pressure.\(^{12}\) In the first few days following closure of a fistula by an operation performed as a physiologic experiment without loss of blood, the increase in total blood volume gradually subsides, since no longer necessary, the abnormally elevated blood pressure also gradually falls, and the retarded heart beat is again elevated to its normal rate.

Following such permanent elimination of a fistula through which blood has flowed with such high velocity and high volume, the regression in the size of the heart and of the palpable proximal artery is astonishingly rapid, coinciding with the remarkably prompt reductions in cardiac output and in total blood volume. The evidence is convincing that the heart and its vessels conform accurately to the volume of blood flowing through them.

In the presence of a peripheral fistula (fig. 1), the components of the circuit of less resistance subjected to greater volume flow and, therefore, to dilatation are the left heart, the aorta, the artery proximal to the fistula, the fistulous opening, the vein proximal to the fistula, the vena cava, the right heart, the pulmonary artery, and the pulmonary vascular bed—in fact the entire circulatory bed except the arterial bed distal to the fistula. The volume of blood sequestered in the fistula circuit, which determines its dilatation, is dependent upon the size of the fistula. Rarely, even the artery distal to the fistula may become dilated by an increased blood volume flow which reaches it and the fistula through a greatly expanded collateral bed.

This occurs when the artery proximal to the fistula is narrowed or obstructed by fibrous tissue contraction, thus preventing it from contributing as much blood as the low resistance fistula is capable of transporting. The capacity of the “hole in the dyke” (the fistula) to transport blood must be satisfied, if not completely by the proximal artery then by added blood by way of an expanding collateral bed and retrograde flow through an expanding artery distal to the fistula (fig. 3).\(^{19, 27}\)

It is important to note that dilatation of the artery proximal to the fistula, which at one time attracted so much attention and so much nebulous thought, is not an isolated process but part of a generalized expansion of all the components of the fistula circuit into which an increased volume of blood is directed through the fistula by the high pressure of the normal circuit.

In the presence of an uncomplicated ventricular septal defect (fig. 4), the components of the circuit of low pressure and low resistance subjected in some cases to a progressively increasing volume flow and, therefore, to progressive dilatation are the left atrium, the left ventricle, the right ventricle, the pulmo-

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**Figure 3**

Diagrammatic presentation (a) of circulatory bed in the presence of a large femoral fistula, the main current of blood entering the fistula by way of the proximal artery, which is dilated; (b) diagrammatic presentation of circulatory bed in the presence of a large femoral fistula, the main current of blood reaching the fistula by way of the dilated collateral circulation and the dilated distal artery, the proximal artery having been ligated.
In the presence of a ventricular septal defect, the shorter circuit in which an increased flow of blood is sequestered, and to which dilatation is limited, consists of the two ventricles, the pulmonary artery, the pulmonary circulatory bed, and the left atrium. The volume of blood sequestered in the circuit of less resistance is determined by the size of the septal defect and by the elasticity or rigidity of the rim of the defect, which determines whether it can or cannot yield to the unrelenting high-pressure thrust of the left ventricle. If rigid and unyielding tissues surround the abnormal orifice, the ventricular septal defect may remain unchanged throughout a comparatively normal life with little or no ventricular dilatation. Some interventricular defects may even recede in size through contracture of the fibrous tissue lining the defect. If the tissues surrounding the septal defect are elastic, the high pressure in the left ventricle will force a progressively increasing volume of blood through the defect into the right ventricle and into the circuit of less resistance, distending and dilating its components, ending eventually in excessive left and right ventricular dilatation and cardiac failure. Should pulmonary vascular sclerosis develop, thus equalizing the pressure in the two ventricles, the flow through the septal defect may not increase and excessive dilatation of the ventricles may not occur.

In the presence of an uncomplicated atrial septal defect (fig. 5), the circuit of low resistance subjected to increased volume flow and, therefore, to dilatation, consists of the left atrium, the right atrium, the right ventricle, the pulmonary artery, the pulmonary vascular bed, and the pulmonary vein. A patient with an uncomplicated atrial septal defect, who was observed before operative closure of the defect was available, lived to age 56 years before fatal right heart failure through excessive dilatation occurred.

In the presence of a patent ductus transporting blood from the aorta to the pulmonary artery (fig. 6), the circuit of low resistance due to the lessened resistance of the pulmonary circulation consists of the left ventricle, the aorta, the patent ductus, the pulmonary artery, the pulmonary vascular bed, the pulmonary vein, and the left atrium. The deflected blood escapes the descending aorta, the systemic peripheral bed, the vena cava, the right atrium, and the right ventricle.
In the presence of a large patent ductus transporting blood from the aorta to the pulmonary artery, the right heart does not share in the increased volume flow, and is therefore not dilated.

Given a ductus with elastic walls capable of yielding ever so minutely but inexorably to the high pressure of the aorta, the flow through the shunt circuit will slowly increase accompanied by a comparably slow progressive dilatation of the components of the shunt circuit. A developing pulmonary vascular sclerosis may effectively limit this increasing flow through the patent ductus and obviate, lessen, or delay the ill effects of dilatation. A patient of 79 years had lived with a ductus for 74 years before symptoms of cardiac failure developed, explicable at autopsy only on the basis of the patent ductus. A 26-year-old patient, with a previously symptomless patent ductus, became pregnant. The increased blood volume and increased cardiac output of pregnancy tipped adversely her previous balance with the patent ductus, which also had produced an increased blood volume and increased cardiac output. Their combined effect caused cardiac over-dilatation and cardiac failure, which required division of the ductus for their control.

If the blood flows through the ductus from pulmonary artery to aorta, as in the fetus and in the rare clinical case (fig. 7), the fistula circuit consists of the right ventricle, the pulmonary artery, the patent ductus, the descending aorta, the systemic peripheral bed, the vena cava and the right atrium, with a smaller volume flow through the pulmonary vascular bed, the pulmonary vein, the left atrium, the left ventricle, and the primary aorta. The right ventricle under these circumstances develops a marked preponderance over the left ventricle due to its abnormally assumed burden of propelling an increased volume of blood through two capillary beds, the resistance in the pulmonary bed equaling or exceeding the resistance in the systemic bed.

Additional abnormal arteriovenous communications, which may occasionally produce similar physiologic and pathologic effects, are rupture of the sinus of Valsalva into the right ventricle; a shunt between the left ventricle and right atrium as reported by Gerbode et al., closure of which produced a tempo-
ry bradycardia, thus indicating an increase in total blood volume due to the shunt; the aortic window in the septum between ascending aorta and pulmonary artery; congenital communications between the coronary artery and sinus; traumatic communications between the internal carotid artery and the cavernous sinus; and the varied congenital hemangiomata, located almost anywhere in the body.

**Summary**

Clinical experiences and experimental studies provide evidence that an abnormal communication between the arterial and venous systems introduces into the circulation two circuits of flowing blood, one characterized by high arterial pressure and high peripheral resistance, and a second system bypassing part of the first, thus introducing a shorter circuit back to the heart, characterized by low pressure and low resistance, each supplied with flowing blood from the same central source, the heart. This shorter circuit acts solely as a parasitic circuit engrafted upon the normal circulation, serving no useful purpose, but capable of producing serious deleterious effects, depending upon the volume of blood diverted from the normal circulation into the shorter or parasitic circuit.

Intimately dependent upon this diversion of differing volumes of blood into the shorter circuit is the great diversity in the effects of a peripheral fistula upon the heart: the complete absence of cardiac dilatation in some cases; the rapid development of cardiac dilatation and cardiac failure within 2½ months as exemplified in Mason's case; and the long delay of 57 years in the development of cardiac failure as observed by Dorney.

Due to the high pressure in the normal circuit and the low resistance in the fistula circuit, there is a constant tendency for blood to be diverted or sequestered, as it were, in the fistulous or parasitic circuit, the factors determining the volume of blood so diverted being the size of the fistula and the distensibility of its rim or border, this, in turn, being dependent upon the character of the fibrous tissue deposited around the fistula in the course of healing. Firm, rigid, and excessive scarring permits only a small volume of blood to be sequestered in the fistula circuit, dilating only mildly the components of the parasitic circuit, whereas an elastic and distensible fistulous border permits progressive expansion of the fistulous opening with a progressive increase in the volume of blood sequestered in the fistula circuit, causing progressive dilatation of all components of this circuit including the four chambers of the heart, the aorta and artery proximal to the fistula, the fistula itself, the proximal vein, the vena cava, and the pulmonary vascular bed.

All gradations in distensibility of the fistula may occur, and hence all gradations in cardiac dilatation will occur. Also, all gradations in the rapidity of fistulous expansion may occur and, therefore, all gradations in rapidity of cardiac dilatation will occur.

This concept of a progressive expansion of a fistula of slow or rapid development permitting more and more blood to be shunted or sequestered in a shorter parasitic circuit at very rapid or very slow rates of increase provides the long-sought explanation of the rapid or long-delayed development of cardiac dilatation and failure in the presence of a peripheral fistula.

It is suggested that the same explanation is applicable to the effects of all abnormal arteriovenous communications, including intracardiac shunts and even some hemangiomata, since all are subject to the same hydrodynamic law that flowing blood like flowing water seeks the path of least resistance.

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Abnormal Arteriovenous Communications: Great Variability of Effects with Particular Reference to Delayed Development of Cardiac Failure

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